Validity and Neuropsychological Characterization of Asperger Syndrome: Convergence with Nonverbal Learning Disabilities Syndrome


Abstract—The authors investigated the validity of Asperger Syndrome (AS) by comparing the neuropsychological profiles in this condition and Higher-Functioning Autism (HFA). Diagnostic assignment followed a stringent procedure based on ICD-10 research criteria for the two disorders. The groups had comparable age and Full Scale IQ distributions. The groups differed significantly in 11 neuropsychological areas. The profile obtained for individuals with AS coincided closely with a cluster of neuropsychological assets and deficits captured by the term nonverbal learning disabilities, suggesting an empirical distinction from HFA.

Keywords: Asperger syndrome, nonverbal learning disabilities, autism

Validity and Neuropsychological Characterization of Asperger Syndrome

Although autism has been widely recognized as the prototypic pervasive developmental disorder (PDD), various other diagnostic concepts with features somewhat similar to autism have been described (Klin & Volkmar, 1995). In contrast to autism, these conditions have been less intensively studied and their validity is more controversial (Volkmar et al., 1994). One of these conditions has been termed Asperger syndrome (AS) (Asperger, 1944), after the Austrian physician who described a number of cases whose clinical features resembled Kanner’s (1943) description of autism (e.g. problems with social interaction and communication, and circumscribed and idiosyncratic patterns of interest). However, Asperger’s description differed from Kanner’s in that speech was less commonly delayed,
motor deficits were more common, the onset appeared to be somewhat later, and all the initial cases occurred in boys (Wing, 1981). Asperger also suggested that similar problems could be observed in family members, particularly fathers.

Asperger’s work was essentially unknown in the English literature for many years. An influential review and series of case reports by Wing (1981) increased interest in AS and since then, both the usage of the term in clinical practice and the number of case reports has been steadily increasing (Gillberg, 1991; Klin, 1994; Szatmari, Tuff, Finlayson & Bartolucci, 1990). The commonly described clinical features of the syndrome include (a) paucity of empathy; (b) naïve, inappropriate, one-sided social interaction, little ability to form friendships, and consequent social isolation; (c) pedantic and poorly intoned speech; (d) poor nonverbal communication; (e) intense absorption in circumscribed topics, such as the weather, facts about TV stations, railway timetables or maps; these are learned in rote fashion and reflect poor understanding, conveying the impression of eccentricity; and (f) clumsy and ill-coordinated movements and odd posture (Wing, 1981). Prevalence rates of 1-10 cases in 10,000 have been suggested (Wing, 1981; Gillberg, 1991), although the lack of generally agreed upon definitions, at least until recently, complicates the interpretation of available research on this point (Tantam, 1988). Although the syndrome was originally reported only in boys, reports of girls with the syndrome have now appeared; it does, however, appear that as in autism, males are significantly more likely to be affected (Szatmari et al., 1990; Volkmar, Szatmari & Sparrow, 1993; Wing, 1991). Although most individuals with the condition function in the normal range of intelligence, some have been reported to be mildly retarded (Wing, 1991). The apparent onset of the condition, or at least its recognition, is probably somewhat later than autism; this may reflect the relatively more preserved language and cognitive abilities (Volkmar & Cohen, 1991). The condition tends to be highly stable (Asperger, 1979), and the higher intellectual skills observed suggest a better long-term outcome than is typically observed in autism (Tantam, 1991).

Validity of Asperger syndrome

The validity of AS as distinct from other conditions, notably the other pervasive developmental disorders, remains controversial (Rutter, 1985; Wing, 1991). Disagreements about the validity of the category and the absence until recently of “official” definitions of AS have meant that the concept has often been used inconsistently by clinicians, who have employed it to refer to autistic persons with higher levels of intelligence, adults with autism, or even as a broader term for all “atypical” children who do not fulfill criteria for autism (Klin, 1994). A lack of uniformity in usage of the term also characterizes definitions adopted for the purpose of research, as different sets of diagnostic criteria have been used by different researchers, with resulting complications for interpretation of research (Ghaziuddin, Tsai & Ghaziuddin, 1992a).

Relative to other pervasive developmental disorders, the validity of AS as distinct from “high functioning autism” (HFA) (i.e. autism associated with overall normal intelligence) has been the topic of greatest debate (Wing, 1991).
There is little disagreement regarding the fact that AS is on a phenomenological continuum with autism, particularly in relation to the problems in the areas of social and communication functioning (Rutter, 1989). For example, within the DSM-III-R diagnostic system, persons with AS would either meet criteria for autistic disorder or would be said to exhibit PDD-NOS (Tsai, 1992). What is less clear is whether the condition is qualitatively different from, rather than just a milder form of, autism unaccompanied by mental retardation (Volkmar & Cohen, 1991). Several studies (Szatmari et al., 1990; Ozonoff, Roge & Pennington, 1991) have attempted to identify discriminating criteria between the two conditions with only mixed results to date. Two factors appear to have contributed to this state of affairs: first, a lack of operationalization and systematic and consensual assignment of the AS diagnosis; and second, a great degree of circularity involved as findings often reflected the criteria adopted in the assignment of a diagnosis of AS and HFA in the first place (Klin, 1994).

On the basis of the available research data, AS was included in both the ICD-10 (WHO, 1990) and DSM-IV (American Psychiatric Association, 1994) diagnostic systems; the two definitions are largely compatible although the ICD-10 system explicitly notes that the validity of the condition apart from HFA remains controversial. However, the advent of the ICD-10 (WHO, 1990) research definition of AS has recently made possible the use of consensual diagnostic criteria. The ICD-10 definition focuses on the differences between AS and autism and takes a more stringent approach than have others adopted by some prominent researchers (Ghaziuddin et al., 1992a), e.g. in ICD-10 the social deficits and unusual behaviors subsumed under the “resistance to change/restricted interest” criteria are the same as for autism but for a diagnosis of AS early language must be near normal.

In the recent DSM-IV Autism/PDD Field Trial (Volkmar et al., 1994), an analysis of cases diagnosed with AS by experienced clinicians yielded some limited evidence for the validity of the condition and its ICD-10 categorical definition. When compared to individuals with HFA (full scale IQ > 85), individuals with AS were less likely to have exhibited delays in the development of spoken language or currently exhibit language/communication deviance; motor delays and “clumsiness” were more variable; isolated special skills (often related to abnormal preoccupations) were more frequent; social, communicative and “resistance to change” symptoms were less frequent; finally, individuals with AS were more likely to exhibit verbal IQ scores greater than performance IQ scores, while the opposite trend was obtained for individuals with HFA. The present study utilizes this recent progress in the nosology of AS and adopts a stringent diagnostic assignment following ICD-10 defining criteria of AS and autism in the investigation of the neuropsychological profiles in these two conditions.

It is also important to note that there are several diagnostic concepts, other than the pervasive developmental disorders, which share, to a great degree, the phenomenological aspects of AS (Bishop, 1989; Denckla, 1983; Rourke, 1989; Voeller, 1986; Weintraub & Mesulam, 1983; Wolff & Chick, 1980). In neuropsychology, a particular subtype of learning disabilities termed the Nonverbal Learning Disabilities syndrome (NLD), has been recently given a great degree of attention (Rourke, 1989). NLD is defined on the basis of a cluster of deficits
affecting the nonverbal aspects of the child's functioning including deficits in tactile perception, psychomotor coordination, visual-spatial organization, nonverbal problem-solving, and appreciation of incongruities and humor. Individuals with the NLD profile are also reported to exhibit well developed rote verbal capacities and verbal memory skills, difficulty in adapting to novel and complex situations and overreliance on rote behaviors in such situations, relative deficits in mechanical arithmetic as compared to proficiencies in single word reading, poor pragmatics and prosody in speech, and significant deficits in social perception, social judgment, and social interaction skills. There are marked deficits in the appreciation of subtle and even fairly obvious nonverbal aspects of communication, which often result in social disdain and rejection by others. As a result, individuals with NLD exhibit a marked tendency toward social withdrawal and are at risk for development of serious mood disorders. Many of the clinical features clustered together in NLD have also been described in the neurological literature as a form of Developmental Learning Disability of the Right Hemisphere (Dencila, 1983; Weintrab & Mesulam, 1983). Children presenting with this condition have also been shown to exhibit profound disturbances in interpretation and expression of affect and other basic interpersonal skills (Voeller, 1986). A familial link has also been suggested (Weintrab & Mesulam, 1983).

Whereas the phenomenological similarities between AS and developmental right-hemisphere dysfunction have been alluded to in the literature (Molina, Ruata & Soler, 1986; Tsai, 1992), NLD has been a relatively unknown concept among psychiatrists (Klin, 1994). The importance of the NLD and related concepts lies in the fact that the neuropsychological profile obtained for individuals so described exhibits a greater right hemisphere dysfunction (Rourke, 1989), whereas the typical neuropsychological profile obtained for individuals with HFA indicates a greater left hemisphere dysfunction (Dawson, 1983; Rumsey, 1992). This would suggest that, based on neuropsychological profiles, there might be at least two types of conditions at the higher end of the spectrum of severe social disabilities: one type with predominantly left hemisphere involvement and the other with right hemisphere involvement (Tsai, 1992). The phenomenological similarities between AS and NLD suggest the use of the NLD profile as a neuropsychological model of AS.

This study was concerned with comparing the neuropsychological profiles of carefully diagnosed individuals with AS and higher-functioning autism (HFA). The neuropsychological profiles obtained for the two groups were hypothesized to differ, with the NLD cluster of assets and disabilities serving as a closer neuropsychological model of AS than HFA. The present exploration of a neuropsychological distinction between the two conditions is the first attempt to explore the validity of AS vis-à-vis HFA while utilizing a stringent application of the ICD-10 defining criteria of AS and autism.

Method

Subjects

Potential subjects (N=73) were recruited from a consecutive case series of individuals seen at the Developmental Disabilities Clinic of our Center and from the membership of the Learning Disabilities
Table 1. Sample Characteristics

<table>
<thead>
<tr>
<th></th>
<th>Age</th>
<th>Sex</th>
<th>Full Scale IQ</th>
</tr>
</thead>
<tbody>
<tr>
<td>HFA (N = 19)</td>
<td>Mean = 15.36 (SD = 9.12)</td>
<td>17 males and 2 females</td>
<td>Mean = 95.63 (SD = 8.27)</td>
</tr>
<tr>
<td>AS (N = 21)</td>
<td>Mean = 16.11 (SD = 8.27)</td>
<td>19 males and 2 females</td>
<td>Mean = 96.76 (SD = 18.19)</td>
</tr>
</tbody>
</table>

N = 40 (24 from Yale, 16 contacted through the LDAA).

Association of America (LDAA). To be eligible for inclusion, each subject had to have extensive records, including developmental and behavioral history and comprehensive neuropsychological data. Of the 73 potential subjects, 24 had been evaluated at the Child Study Center while the remaining 49 had undergone similar evaluations elsewhere. Inclusionary criteria included a Full Scale IQ of over 70 as well as a diagnosis of AS or autism according to both experienced clinicians (FV and AK) and ICD-10 research criteria.

From the potential subject pool, 33 cases were eliminated from subsequent analysis because either one or more Full Scale IQ scores of below 70 had been obtained in the individual’s lifetime or, more commonly, because a diagnosis of either AS or HFA could not be unequivocally confirmed. The remaining subjects, 21 with AS and 19 with HFA, constituted the final sample. The groups did not significantly differ in terms of age, sex, and Full Scale IQ distributions. A summary of sample characteristics is given in Table 1.

Diagnostic assignment. Psychiatric diagnosis was assigned without knowledge of the subjects’ neuropsychological characterization (see below), after a thorough reviews of records by two of the authors (AK and FRV) who also completed the ICD-10 rating criteria for autism and AS (see Table 2). As noted above, potential subjects for whom there was no concordance between clinician-assigned diagnosis and ICD-10 ratings were excluded from the study. To ensure that the diagnosis of AS was made on the strictest possible basis, the draft ICD-10 research definition was modified in some respects. Firstly, consistent with the results of the DSM-IV Autism/PDD field trial (Volkmar et al., 1994), only one item in the symptom cluster of “restricted interests and activities” was required, and (2) the associated features in ICD-10 of “delayed motor milestones and presence of motor ‘clumsiness’,” and “isolated, unusual all-absorbing skill or activity”, that are considered suggestive (but not required) for an ICD-10 diagnosis of AS, were necessary diagnostic criteria. This most stringent approach appeared to us to be most consistent with the particularly distinctive features of AS as described in the literature (Klin, 1994).

Table 2. Defining criteria for HFA and AS

**Childhood Autism** (ICD-10 DRAFT Research Criteria)

1. 1 out of 3 onset criteria
2. 3 out of 5 criteria involving “qualitative impairments in reciprocal social interaction”
3. 2 out of 5 criteria involving “qualitative impairments in communication”
4. 2 out of 6 criteria involving “restricted, repetitive, and stereotyped patterns of behavior, interests and activities”
5. Exclusion of other disorders

**Asperger Syndrome** (ICD-10 DRAFT Research Criteria)

1. “A lack of clinically significant general delay in language or cognitive development” + onset criteria
2. “Qualitative impairments in reciprocal social interaction” (criteria as for autism)
3. “Restricted, repetitive, and stereotyped patterns of behavior, interests and activities” (criteria as for autism) + 1 endorsed item sufficient (rather than the required 2)
4. Exclusion of other disorders (e.g. autism, PDD-NOS)
5. Delayed motor milestones and presence of motor “clumsiness”
6. Isolated, unusual all-absorbing special skill or activity
Table 3. Defining criteria for Nonverbal Learning Disabilities (NLD)

<table>
<thead>
<tr>
<th>Assets</th>
<th>Deficits</th>
</tr>
</thead>
<tbody>
<tr>
<td>Auditory perception</td>
<td>Fine motor skills</td>
</tr>
<tr>
<td>Rote material</td>
<td>Gross motor skills</td>
</tr>
<tr>
<td>Auditory/verbal memory</td>
<td>Visual-motor integration</td>
</tr>
<tr>
<td>Phonology</td>
<td>Visual-spatial perception</td>
</tr>
<tr>
<td>Vocabulary</td>
<td>Novel material</td>
</tr>
<tr>
<td>Verbal output</td>
<td>Visual memory</td>
</tr>
<tr>
<td>Word decoding/spelling</td>
<td>Verbal concept formation</td>
</tr>
<tr>
<td></td>
<td>Nonverbal concept formation</td>
</tr>
<tr>
<td></td>
<td>Prosody</td>
</tr>
<tr>
<td></td>
<td>Verbal content</td>
</tr>
<tr>
<td></td>
<td>Pragmatics</td>
</tr>
<tr>
<td></td>
<td>Reading comprehension</td>
</tr>
<tr>
<td></td>
<td>Mechanical arithmetic</td>
</tr>
<tr>
<td></td>
<td>Social competence</td>
</tr>
<tr>
<td></td>
<td>Emotional competence</td>
</tr>
</tbody>
</table>

The interrater reliability for both clinician-assigned diagnosis and ICD-10 rating criteria were examined by the random selection of half of the sample (i.e. 20 subjects) and having their records reviewed and their diagnostic procedure completed by an additional experienced clinician unaware of previous diagnostic assignment. The interrater reliability, as measured by the chance-corrected coefficient Kappa (K) and proportion of observed agreement (PO) (Siegel & Castellan, 1988) was excellent for clinician-assigned diagnosis (K = .799, SE = .095; PO = .90), and fair to excellent for the diagnosis criteria (range of K .50-.80; range of PO .75-.95) (Cicchetti & Sparrow, 1981).

Neuropsychological characterization. Twenty-two items (see Table 3), including seven areas of assets and 15 areas of deficits that define the condition of Nonverbal Learning Disabilities (NLD) (Rourke, 1989) were rated by an experienced neuropsychologist on the basis of a thorough review of neuropsychological records. These criteria were rated in terms of assets or deficits, depending on whether results in normed instruments or clinical observations were described as relative assets or clinically significant deficits in the various areas examined. A conservative algorithm for the NLD+ or NLD− characterization was defined, with the former requiring the concordance of at least 10 of the 15 deficits and five of the seven assets. Deficits in tactile perception were excluded from this rating system because of the low frequency with which such deficits were mentioned in the reports of neuropsychological assessments. It should be noted that the following items were rated on a basis of, mostly, clinical observations rather than normed instruments: Gross Motor skills (as observed or measured during the evaluation, rather than reported on the basis of the subject’s “clumsiness” in real-life situations, e.g. performance at sports or use of equipment requiring a minimal degree of motor coordination); phonology (i.e. articulation), and prosody (primarily intonation and modulation of volume, although, in some cases, also pitch and rhythm); verbal output (frequency and length of utterances), verbal content (coherence of utterances and ability to convey logical thoughts), pragmatics (e.g. relevance and quantity of utterances, reciprocity, topic management—i.e. demarcation of beginning and ending of topic), social competence (e.g. understanding and conformity to rules and conventions of social interaction, friendships, and so forth), and emotional competence (e.g. empathy, affect modulation).

The interrater reliability of ratings of neuropsychological criteria was examined by randomly selecting one-half of the sample (i.e. 20 subjects) and having their neuropsychological records reviewed and their ratings completed by an additional neuropsychologist. The interrater reliability for the NLD+/NLD− characterization was very good (K = .749, SE = .105; PO = .90), and fair to excellent for the rating criteria (range of K .48-.80; PO .75-1.00) (Cicchetti & Sparrow, 1981).
### Table 4. Differential patterns of verbal IQ, performance IQ, and verbal–performance IQ discrepancy for the HFA and AS groups

<table>
<thead>
<tr>
<th></th>
<th>HFA Mean (SD)</th>
<th>AS Mean (SD)</th>
<th>Statistical significance</th>
</tr>
</thead>
<tbody>
<tr>
<td>Full Scale IQ</td>
<td>95.63 (14.66)</td>
<td>96.76 (18.19)</td>
<td>NS</td>
</tr>
<tr>
<td>Verbal IQ (VIQ)</td>
<td>94.63 (22.53)</td>
<td>108.95 (18.52)</td>
<td>p &lt; .001</td>
</tr>
<tr>
<td>Performance IQ (PIQ)</td>
<td>96.68 (12.56)</td>
<td>85.14 (18.81)</td>
<td>p &lt; .01</td>
</tr>
<tr>
<td>VIQ – PIQ</td>
<td>-2.05 (25.69)</td>
<td>+23.81 (14.07)</td>
<td>p &lt; .001</td>
</tr>
</tbody>
</table>

### Procedures

The neuropsychological data for the AS and HFA groups were compared in terms of Verbal IQ—Performance IQ difference, overlap between psychiatric diagnosis (i.e. AS and autism), neuropsychological characterization (i.e. NLD+/NLD−), and presence of deficits in the 22 neuropsychological areas defining the NLD syndrome.

### Results

Although the AS and HFA groups did not differ in terms of Full Scale IQ, the Verbal and Performance IQ (VIQ and PIQ), and particularly the VIQ–PIQ differential (i.e. VIQ–PIQ) were significantly different. The AS group exhibited a higher VIQ and lower PIQ in comparison to the HFA group (see Table 4). The IQ distribution of the AS group indicates that, for this sample, a higher VIQ than PIQ was a universal finding. A repeated measures analysis of variance of clinical group (i.e. AS and HFA) by IQ type (i.e. verbal and performance) revealed a significant interaction between clinical diagnosis and IQ type ($F_{1,38} = 16.05$, $p < .001$).

When the degree of overlap between psychiatric diagnosis (i.e. AS/HFA) and neuropsychological characterization (i.e. NLD+/NLD−) was examined, a high degree of concordance between AS and NLD+ was observed. This suggests that the NLD profile was indeed an adequate model of neuropsychological assets and deficits encountered in individuals with AS. As can be seen in Table 5, this was not the case for individuals with HFA.

In order to compare the neuropsychological profile of the two groups in more detail, the frequency of subjects exhibiting deficits on the 22 areas defining the NLD characterization was examined. As can be seen in Table 6, 11 of the 22 items

### Table 5. Overlap between psychiatric diagnoses (HFA and AS) and neuropsychological characterization (NLD+ and NLD−)

<table>
<thead>
<tr>
<th></th>
<th>NLD+</th>
<th>NLD−</th>
</tr>
</thead>
<tbody>
<tr>
<td>HFA</td>
<td>1</td>
<td>18</td>
</tr>
<tr>
<td>AS</td>
<td>18</td>
<td>3</td>
</tr>
</tbody>
</table>

Fisher’s exact $p = .000$ ($p < .001$).
Table 6. Frequency of subjects exhibiting deficits on the 22 NLD items

<table>
<thead>
<tr>
<th>Item</th>
<th>HFA</th>
<th>AS</th>
<th>Fisher’s $p$</th>
</tr>
</thead>
<tbody>
<tr>
<td>Fine motor skills</td>
<td>6/19</td>
<td>19/21</td>
<td>.001 ***</td>
</tr>
<tr>
<td>Gross motor skills</td>
<td>12/19</td>
<td>21/21</td>
<td>.003 **</td>
</tr>
<tr>
<td>Visual-motor integration</td>
<td>8/19</td>
<td>19/21</td>
<td>.002 **</td>
</tr>
<tr>
<td>Visual-spatial perception</td>
<td>5/19</td>
<td>16/21</td>
<td>.004 **</td>
</tr>
<tr>
<td>Auditory perception</td>
<td>10/19</td>
<td>1/21</td>
<td>.001 **</td>
</tr>
<tr>
<td>Novel material</td>
<td>11/19</td>
<td>15/21</td>
<td>.510</td>
</tr>
<tr>
<td>Rote material</td>
<td>2/19</td>
<td>0/21</td>
<td>.219</td>
</tr>
<tr>
<td>Verbal memory</td>
<td>11/19</td>
<td>5/21</td>
<td>.049 *</td>
</tr>
<tr>
<td>Visual memory</td>
<td>9/19</td>
<td>19/21</td>
<td>.005 **</td>
</tr>
<tr>
<td>Verbal concept formation</td>
<td>10/19</td>
<td>10/21</td>
<td>1.000</td>
</tr>
<tr>
<td>Nonverbal concept formation</td>
<td>5/19</td>
<td>16/21</td>
<td>.004 **</td>
</tr>
<tr>
<td>Articulation</td>
<td>11/19</td>
<td>1/21</td>
<td>.001 ***</td>
</tr>
<tr>
<td>Vocabulary</td>
<td>8/19</td>
<td>0/21</td>
<td>.001 **</td>
</tr>
<tr>
<td>Verbal output</td>
<td>11/19</td>
<td>1/21</td>
<td>.001 ***</td>
</tr>
<tr>
<td>Verbal content</td>
<td>18/19</td>
<td>20/21</td>
<td>1.000</td>
</tr>
<tr>
<td>Prosody</td>
<td>17/19</td>
<td>19/21</td>
<td>1.000</td>
</tr>
<tr>
<td>Pragmatics</td>
<td>16/19</td>
<td>20/21</td>
<td>.331</td>
</tr>
<tr>
<td>Word decoding</td>
<td>7/19</td>
<td>2/21</td>
<td>.060</td>
</tr>
<tr>
<td>Reading/comprehension</td>
<td>10/19</td>
<td>10/21</td>
<td>1.000</td>
</tr>
<tr>
<td>Arithmetic</td>
<td>8/19</td>
<td>9/21</td>
<td>1.000</td>
</tr>
<tr>
<td>Social competence</td>
<td>19/19</td>
<td>21/21</td>
<td>1.000</td>
</tr>
<tr>
<td>Emotional</td>
<td>19/19</td>
<td>21/21</td>
<td>1.000</td>
</tr>
</tbody>
</table>

*p < .05; **p < .01; ***p < .001.

discriminated between the AS and HFA groups. Of these 11 items, six were found
to be predictive of a diagnosis of AS, whereas five were predictive of a diagnosis
of “Not–AS” (Table 7).

It should be noted that four areas were found to be deficient for the great
majority of subjects in both AS and HFA groups: three of these (“prosody”, “social
and emotional competence”) are subsumed under the diagnostic definition of AS
and autism, whereas one (“verbal content”) is not. Of the remaining 18 areas, only
deficits in “Gross Motor Skills” and “Fine Motor Skills” could possibly be subsumed
under the diagnostic criterion for AS “motor clumsiness”, although the latter has
rarely been operationally defined or systematically described (Ghaziuddin, Tsai &
Ghaziuddin, 1992b). Therefore, with the possible exception of five items (gross
and fine motor skills, prosody, and social and emotional competence), the remaining
areas of neuropsychological characterization appeared to be independent of the
initial psychiatric diagnostic assignment. Of these five items, three areas (prosody
and social and emotional competence) did not differ between the AS and HFA
groups.

Finally, as shown in Table 3, the operational definition of NLD+ was made in
terms of an a priori decision regarding the required number of concordant assets
and deficits as predicted by the NLD model (i.e. ≥5 out of 7 assets, and ≥10 out
of 15 deficits, see Table 3. As this decision had been made on a theoretical basis
prior to data collection, we explored the utility of this cut off point for a prediction
of a diagnosis of AS in the case of the present sample of individuals with AS and HFA. However, the adopted cut off point was only one of 128 possible cut off points (i.e. 0–7 out of 7 assets, AND 0–15 out of 15 deficits, hence $8 \times 16 = 128$). In order to obtain an empirically-derived optimal cut off point, it was necessary to analyze the utility of the a priori cut off point vis-à-vis all other possible cut off points. The sensitivity, specificity and coefficient of agreement Kappa for all 128 combinations were calculated (Kraemer, 1988). Eight combinations were associated with a Kappa $\geq .75$ (“excellent” agreement (Cicchetti & Sparrow, 1981). The sensitivity and specificity for these combinations ranged from .76 to .90 and .95 to 1.00, respectively. The a priori combination (i.e. $\geq 5$ out of 7 assets, and $\geq 10$ out of 15 deficits) exhibited a sensitivity of .86, a specificity of .95, and a Kappa of .80. The highest Kappa (.85) was exhibited by the combination including $\geq 3$ out of 7 assets, and $\geq 10$ out of 15 deficits (sensitivity of .90 and specificity of .95). These results suggest that (a) the a priori decision regarding the operational definition of NLD+ proved to be highly predictive of a diagnosis of AS in the present sample; and (b) as a number of variations of the NLD+ operationalization were highly predictive of a diagnosis of AS in the present sample, the present results also suggest that the overlap between NLD+ and AS is a rather robust phenomenon.

Discussion

This study examined the neuropsychological profiles of a group of AS and HFA individuals whose diagnostic assignment was made of following stringent criteria
and an algorithm based on ICD-10 research criteria for the two conditions. A comparison of neuropsychological profiles revealed that 11 areas discriminated between the two conditions, nine of which were apparently independent from the psychiatric diagnostic assignment. The high level of concordance between AS (but not HFA) and a neuropsychological characterization of NLD suggests that the latter can be seen as an adequate neuropsychological marker of AS. Interestingly, such neuropsychological differences between AS and HFA were captured in the groups’ VIQ-PIQ discrepancy, with VIQ being universally higher than PIQ for individuals with AS, whereas no differential was found for individuals with HFA, a finding that is supported by results of the DSM-IV Autism/PDD field trial (Volkmar et al., 1994) and recent neuropsychological research of autism unaccompanied by mental retardation (Minshew, 1992).

The present findings contrast with previous work on the neuropsychology of AS (e.g. Ozonoff et al., 1991; Szatmari et al., 1990). However, the present use of stringent ICD-10 criteria for diagnostic assignment complicates a direct interpretation of differences with previous studies where less systematic and stringent diagnostic assignment was adopted. For example, Szatmari et al.’s study (1990) revealed no significant differences between AS and HFA groups, leading the authors to the conclusion that “it is justified to combine the AS and HFA groups into a more general PDD category (p. 135). However, the adopted inclusionary criteria—(1) isolated behavior; (2) impaired social interaction; (3) one of odd speech, impaired nonverbal communication, or bizarre preoccupations, and (4) onset prior to 6 years of age (p. 131)—defined AS in a much broader fashion than the ICD-10 draft research criteria. The authors’ conclusion that AS appears to be “a very mild form of PDD” (p. 136) raises some questions as to the face value of creating the AS grouping apart from the PDD-NOS category, and may account for some of the positive findings (with regard to early history), and the rather broad definition of the syndrome may partly account for the lack of findings in regard to the groups’ neurocognitive profiles. Clearly, research findings are dependent upon the nosologic approach adopted. In contrast to Szatmari et al.’s strategy, the present study adopted the ICD-10 approach to AS, which attempts to maximize the differences between HFA and AS in ways compatible with, as yet tentative, clinical research data.

Ozonoff et al.’s study (1991) did use the ICD-10 definition of AS, but modified it by excluding the onset criteria. The exclusion was justified in terms of the current debate regarding the validity of the AS onset criteria (Gillberg & Steffenburg, 1987; Wing, 1988). Although this approach could be well justified, it did make the AS definition broader than the one adopted in the present study. The impact of a given diagnostic system on the assignment of diagnosis is illustrated in Ozonoff et al.’s (1991) finding that 40% of their AS subjects met DSM-III-R (1987) criteria for autism. In many respects, Ozonoff et al.’s (1991) findings were inconsistent with Szatmari et al.’s (1990). For example, in Ozonoff et al.’s (1991) study, the AS and HFA groups did significantly differ in terms of Verbal IQ—Performance IQ differential due to the AS group having significantly higher Verbal IQ.

Clearly, the nosological strategy has a direct impact on findings of studies of AS. Given the current state of affairs, our approach was to select the most
phenomenologically prototypical cases of HFA and AS, with a view to maximize differences in developmental history and presentation. Operationally, we decided to adopt the most stringent current definitions of these syndromes. The rationale for this strategy was based on the assumption that if neuropsychological differences could not be found with the extreme cases, then there would be little reason to pursue this line of research. In this effort, we chose to add two additional criteria, which are suggested but not required in ICD-10 and DSM-IV (American Psychiatric Association, 1994), so as to further specify our definition of AS. The two criteria were (a) "delayed motor milestones and presence of motor 'clumsiness'", and (b) "isolated, unusual all-absorbing special skill or activity". This strategy entailed some gains and losses. More stringency was achieved by bringing our defining criteria closer to findings revealed in the Autism/PDD DSM-IV field trial (Volkmar et al., 1994), according to which it was the case that motor delays and clumsiness, and idiosyncratic/circumscribed interests were very common in AS. However, this approach entailed a modification of the ICD-10 defining criteria, and this should be taken into consideration when interpreting our findings.

For example, the introduction of the motor criterion may have selected a very specific subsample of AS subjects from our original pool of potential participants. This possibility prompted a reanalysis of our data with a view to ascertain whether any additional subjects would have been included in the AS sample had we not made the motor criterion necessary for the diagnosis. From the 33 individuals excluded, only six subjects would have been included in the AS sample using this less restrictive definition; interestingly, however, all these were complex cases which were excluded from further analysis owing to a lack of concordance between clinician-assigned diagnosis and the ICD-10 rating criteria. The inclusion of these six subjects in the AS sample did not significantly alter the results obtained, although the statistical significance level of group comparisons for four neuropsychological items—gross motor skills, visual-motor integration, visual-spatial perception, and visual memory—dropped slightly. There was only one significant alteration of results, and that concerned the group comparison for verbal memory, which became barely significant at the \( p < .05 \) level. Paradoxically, this was one area for which there was significant differences between the AS and HFA groups in Ozonoff et al.'s (1991) study. This reanalysis notwithstanding, it is still possible that our AS sample represented a specific group of AS individuals whose neuropsychological profile includes motor symptoms.

Another issue relates to the ICD-10 (and our) requirement for "preservation" of language skills early in development, and its possible relationship to later verbal/nonverbal IQ differences as a result of a "downstream" effect. Following this reasoning, the group differences for verbal/performance IQ differential would simply reflect higher verbal skills from the onset. Although this is in fact an empirical issue, it should be clarified that the ICD-10 meaning of "lack of clinically significant delays or deviance in language acquisition" relates to early language, and not language at the time of our examination (as adopted in Ozonoff et. al.'s (1991) study). As for the possible "downstream" effect, it is indeed possible that later differences may reflect a more basic initial difference in language development. However, the issue of early language acquisition should be seen as rather separate
from later language/communication skills. The early preservation of language
does not *necessarily* imply that later language skills will be normal; in fact, they often
appeared not to be normal, particularly in terms of the suprasegmental, pragmatic,
and competence (e.g. nonliteral) aspects of language. That notwithstanding,
certain aspects of language functioning (particularly the formal aspects, e.g.
vocabulary, grammatical competence) did appear to represent continued areas of
strength for individuals with AS.

A final point in regard to the interpretation of our findings concerns the
recruitment origin of our sample. In order to ascertain any differences between
the Yale group (N = 24) and the LDAA group (N = 16), we compared the two
samples for age, gender, and Full Scale IQ. Although there were no significant
differences in terms of age and Full Scale IQ, there were differences in terms of
gender, as both female subjects with AS were from the LDAA group.

In summary, the various issues arising from the adoption of different diagnostic
approaches in research studies serve to underscore the paramount importance of
stringent nosology for the validation and characterization of diagnostic concepts
that share several features such as AS and HFA.

The present findings can be seen as a contribution to the validation of AS vis-
'à-vis HFA, as these two conditions could be distinguished in terms of their
neuropsychological characterization. This is a more complex issue, however, given
that these two conditions could still share the same etiology or other pathogenetic
processes while having phenotypic difference solely accounted for by
neuropsychological differences. In this sense, AS and HFA could be seen as the
same diagnostic entity expressed differently because of different neuropsychological
endowment, similar, to some extent, to the phenomenological differences between
lower functioning and higher functioning autism. Whether AS and HFA are truly
different diagnostic entities will be clarified only with additional research on the
developmental, behavioral genetics, and neuroanatomic aspects of these two
conditions. That notwithstanding, the convergence between AS and NLD suggests
lines of investigation focused on right-hemisphere abnormalities, or other competing
neuroanatomic models [e.g. involving white matter abnormalities (Rourke, 1989)]
purported to account for the unique cluster of neuropsychological assets and
deficits characterizing NLD. Such research may review mechanisms responsible
for disruptions of socialization skills of a different nature than the ones still eluding
investigators of autism.

The clarity of results obtained in this study may have been a function of the
investigators' decision to include only stringently diagnosed, prototypical cases of
AS and HFA. The phenotypic expression of both conditions is probably more
heterogenous than evidenced in the present sample (Klin & Volkmar, 1995).
However, given the current controversy regarding the validity of AS, our results
suggest the importance of utilizing a stringent and operational definition of subjects
studied. Conversely, it will be important to explore the possible associations between
neuropsychological profiles and varied clinical presentations of this and other
syndromes characterized by disabilities of socialization (e.g. the study of individuals
with a neuropsychological profile consistent with NLD, but who do not meet criteria
for a pervasive developmental disorder).
Finally, regardless of whether or not AS and HFA are truly different diagnostic entities, the significant divergence of neuropsychological profiles suggests that intervention strategies for AS should be of a different nature, directly addressing specific neuropsychological deficits and building on neuropsychological assets, an approach that has been described as very useful with individuals with Nonverbal Learning Disabilities (Klin, 1994; Rourke, 1989).

References


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