Asperger's Syndrome: A Comparison of Clinical Diagnoses and Those Made According to the ICD-10 and DSM-IV

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The diagnostic criteria for Asperger Syndrome (AS) according to ICD-10 and DSM-IV have been criticized as being too narrow in view of the rules of onset and precedence, whereby autism takes precedence over AS in a diagnostic hierarchy. In order to investigate this further, cases from the DSM-IV multicenter study who had been diagnosed clinically with AS were assigned to appropriate DSM-IV/ICD-10 diagnostic categories. The analysis indicated that 11(23%) cases would be reassigned a diagnosis of autism by either ICD-10 or DSM-IV according to their onset and precedence rules, and 33(68%) would be diagnosed with AS. These results contrast with those of others who have stated that the diagnosis of AS using ICD-10/DSM-IV criteria is 'virtually impossible'. It is suggested that this is due to limitations inherent in these criteria, and alternative conceptualizations are discussed.

KEY WORDS: Asperger syndrome; diagnosis; autism.

Current conceptualizations of autistic disorder (AD) and Asperger Syndrome (AS) have evolved from the original clinical descriptions of relatively small numbers of cases of children by Kanner (Kanner, 1943) and Asperger (Asperger, 1944, translated in Frith, 1991), respectively (see Klin & Volkmar, 1997 for a review). Diagnostic criteria, in terms of what characteristics designate 'caseness', were not made explicit by either clinician. This has subsequently lead to confusion regarding the conceptual boundaries of the two disorders and a lack of understanding of the nature of the relationship between them, despite an abundance of research devoted to addressing these issues (see Volkmar &

Whilst AD was first included in both ICD-9 and DSM-III (American Psychiatric Association (APA), 1987), it was not until the most recent volumes, i.e. ICD-10 (World Health Organization (WHO), 1992) and DSM-IV (APA, 1994), that AS was included (termed Asperger Disorder in DSM-IV). Asperger (1944/1991) had originally described a group of older children who, although socially motivated, had

Klin, 2000 for a discussion of diagnostic issues, and Ozonoff, 2000 for a discussion of the neuropsychological characteristics of the two disorders). Moreover, the relationship between these disorders and other diagnostic labels assigned to people with difficulties in social interaction, and which have evolved from different areas of clinical expertise, such as non-verbal learning disability (NLD) from neuropsychology (Rourke, 1987, 1989), semantic-pragmatic disorder from psycholinguistics (Bishop, 1989), and schizoid personality from adult psychiatry (Wolff, 1991; Wolff & Barlow, 1979), is equally poorly understood (also see relevant chapters in Klin, Volkmar, & Sparrow, 2000 for up to date discussion of these diagnostic concepts).

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difficulties engaging others because of their difficulty reading non-verbal social cues, a lack of understanding of conversational rules, and a tendency to talk at length about particular topics of interest about which they knew a great deal, and the pursual of which would occupy much of their time to the detriment of forming relationships with others. Asperger believed there was a fundamental difference between the syndrome he described and that of Kanner, emphasizing its apparently later onset, and although the relationship between the two is still the focus of much debate, genetic, neuropsychological and outcome studies appear to support a relationship between the two. This has lead some researchers to argue for the existence of an autistic spectrum (Wing, 1998), and the suggestion that the term AS, synonymous with higher cognitively functioning autism, is redundant (Schopler, 1985).

Even before AS was included in the ICD-10 and DSM-IV, clinicians were making clinical diagnoses of AS, based on the descriptions provided by Asperger (1944/1991), and subsequently by Wing (1981), who had introduced Asperger's syndrome to the English Language scientific community in a description of a series of cases resembling those first described by Asperger (Wing, 1981). During the decade of the 1980s several other clinicians attempted to define the core characteristics of AS, and created operationalized criteria (Gillberg & Gillberg, 1989; Tantam, 1988). Unfortunately, this increased the confusion, as different clinicians began to diagnose according to different criteria. Moreover, this was compounded by the concept being used in yet other ways by other clinicians: for example, as a mild form of autism (Gillberg & Gillberg, 1989), as a synonym for PDDNOS (Szatmari, Bremner, & Nagy, 1989), or to describe higher functioning people with autism, creating a so-called 'autistic spectrum' (Wing, 1997; see Volkmar & Klin, 2000 for a discussion). The inclusion of AS into the ICD-10 and subsequently the DSM-IV was an attempt to reduce this nosologic heterogeneity and provide a consistent set of criteria that would allow important questions regarding the validity of AS to be investigated.

The diagnostic criteria for AS in both ICD-10 and DSM-IV include qualitative abnormalities in reciprocal social interaction (criteria as for autism), and restricted and repetitive stereotyped patterns of behavior (as for autism). The disorder also includes specific onset criteria, namely that there is no history of significant general delay in spoken language, and that self-help skills, adaptive behavior and curiosity

about the environment should be at a level consistent with normal development. In addition, autism takes precedence in the hierarchy of diagnosis: if the person meets the diagnostic criteria for autism, i.e. has evidence of the presence of early developmental abnormalities, and also has impairments defined by the communication domain on the autism criteria, this takes precedence. As noted subsequently this definition was influenced by the results of a large multi-cite field trial (Volkmar, Klin, Siegel, Szatmari, et al., 1994).

Whilst the development of diagnostic criteria has alleviated inconsistencies to a certain extent, there are still some major problems. In particular, they have been criticized as being too narrow, in view of the 'precedence' and the 'onset' criteria described above, to the extent that assigning people to this category is unlikely (Eisenmajer et al., 1996; Mayes, Calhoun, & Crites, 2001; Miller & Ozonoff, 1997). Not all studies have found this to be the case. however. Hippler and Klicpera (2003), in a reanalysis of 44 cases diagnosed by Asperger with 'autistic psychopathy', demonstrated that 30 (68%) would be diagnosed according to ICD-10 criteria (Hippler & Klicpera, 2003). Other criticisms have related to the failure to include additional features described by Asperger. For example, the presence of motor clumsiness was noted by Asperger (1944/1991), and subsequent clinicians who described the core characteristics (Gillberg & Gillberg, 1989; Tantam, 1988; Wing, 1981), but is not included, and communication impairments, involving the so-called pragmatic aspects of language rather than semantics or syntax, are also not included (for an in-depth review see Landa, 2000).

The current reanalysis of the DSM-IV multicenter field trial data was undertaken to determine the extent to which people who were clinically diagnosed with AS before the introduction of either the ICD-10 or DSM-IV would meet the inclusion criteria. It was hypothesized that many of the people who had been clinically diagnosed with AS would not meet the ICD-10 or DSM-IV criteria, in view of the precedence and early onset criteria. It was also hypothesized that some of these people would in addition fail to meet the criteria for autistic disorder, in view of the communication criteria. Finally, the prevalence of two additional criteria originally described by Asperger not explicitly included in either DSM-IV or ICD-10 were considered: namely, motor clumsiness and special skills.

METHODS

Cases

The DSM-IV multicenter field trial, a collaborative study based at more than twenty sites around the world, was undertaken to inform the development of the DSM-IV criteria for autistic disorder. The results of the Field Trial are available in a report and previous scientific papers where a more detailed account of the methodology, in terms of case identification and procedures undertaken, can be found (Volkmar, Klin, Siegel, Szatmari, & et al., 1994). In short, 977 cases, of whom 48 had been diagnosed with AS, were identified by the collaborative centers. Although the Field Trial was not primarily designed to evaluate the validity of the diagnostic category of AS, cases had been specifically recruited from certain centers for the purpose of ensuring adequate representation of cases that might exhibit AS. A standard coding system was used to provide subject background details, such as gender, ethnicity, age and IQ, and to provide information regarding item scores for each subject for the various diagnostic criteria considered, namely DSM-III, DSM-IIIR and ICD-10.

Analysis

For the present study, only those participants with a clinical diagnosis of AS are considered. From the complete database, in addition to background characteristics, the following information was retained for analysis: first, whether each person met the diagnostic criteria for the 'social impairment' and 'repetitive patterns of behavior' domains for AD/AS, and 'communication' domain for autism; secondly, information relating to early history of developmental delays of language, cognition, and adaptive behavior; thirdly, early history of presence of special abilities; and fourthly, early history of clumsiness. Each participant was coded according to whether or not he/she met the diagnostic criteria for AS defined according to ICD-10 or DSM-IV. This involved first considering whether they met the criteria for autism (the precedence rule), and second, whether they met the onset criteria (the onset rule). As the diagnostic criteria for both systems are virtually identical they are not considered separately.

RESULTS

The 48 individuals with a clinical diagnosis of AS included 38 male and 10 female cases, with a mean

age 12 years 7 months (SD 8 years 1 month), and mean FSIQ 95.9 (SD 19.6).

Consistent with the original hypothesis, a significant number of cases were reassigned to the category of autistic disorder (AD) in DSM-IV and childhood autism in ICD-10. This included 11 cases (23%) who had evidence of impairments in all three developmental domains, with additional evidence of delay in the acquisition of language. Contrary to the original hypothesis, however, the majority of the clinical cases of AS were diagnosed with AS according to both ICD-10 and DSM-IV. These cases were made up of two groups. The first group (N=18, 38%) included those who had impairments in all three developmental domains, but without any early history of delay in language or cognitive development. The second group (N=15, 31%) consisted of those who had impairments in the two domains of social interaction and restricted and repetitive patterns of behavior without any history of delay in language or cognitive development. The remaining cases included four participants (9%) who were impaired in the two domains of social interaction and restricted and repetitive patterns of behavior but who had evidence that early language/cognitive development was delayed. The ICD-10 and DSM-IV would assign such individuals to the 'other PDD' and 'PDDNOS' categories respectively.

Next, the prevalence of other behaviors described by Asperger but not specifically included in the diagnostic criteria (although mentioned) were considered, namely motor clumsiness and the presence of 'special skills'. Of the 48 cases, 23 (48%) were described as exhibiting motor clumsiness, and 37 (77%) as having special skills. When the group preferentially described as AD was compared with the group described as AS according to the two diagnostic systems, no differences were observed for intellectual ability, presence of motor clumsiness or special skills.

DISCUSSION

Although Asperger Syndrome was included as a diagnosis in the ICD-10 and DSM-IV, following recognition that people with a clinical diagnosis could reliably be differentiated from high functioning autism, it has been argued that the conceptualization is too narrow, to the extent that assigning the diagnosis is almost impossible as a result of the 'precedence' and 'early onset' rules. The current study

examined the applicability of the ICD-10 and DSM-IV criteria to 48 individuals diagnosed clinically with AS independent of either nosological system. The results indicated that although a significant number of people with a clinical diagnosis were reassigned a diagnosis of autism as a result of these rules, this was not true for the majority of cases, whose clinical diagnosis of AS was consistent with the two sets of criteria. Although this result contrasts with several other papers that have criticized these criteria as being too narrow (Leekam, Libby, Wing, Gould, & Gillberg, 2000; Mayes, Calhoun, & Crites, 2001; Miller & Ozonoff, 1997), they reflect those of Hippler and Klipera (2003), who reanalyzed the cases of Asperger according to the ICD-10 criteria and found that a significant majority would still fulfill the diagnosis of AS even when the precedence rule and early onset criteria are considered. Two other possible markers, motor clumsiness and special skills, although demonstrated in a significant number of those with a clinical diagnosis of AS, were not demonstrated in the majority of cases and the same was true of special skills. Moreover, these did not differentiate between the final ICD-10/DSM-IV groups.

Although on one level these results support the conceptualization of two disorders of social disability, one of early onset and characterized by social difficulties and rigid patterns of behavior with communication impairments, and the other of later onset and characterized by similar social and behavioral impairments, there are several issues with the criteria as presently defined, which help to explain why some researchers have found lack of consonance between the clinical and DSM-IV/ICD-10 concepts, whilst others have not. In particular, using onset as an inclusion/exclusion criteria, whereby individuals are considered for a diagnosis of AS only in the absence of early speech delay or impairments of self-help skills, adaptive behavior or curiosity about the environment, has several disadvantages. A fundamental concern is that this tends to 'tilt' the diagnosis towards autism on the basis of vague developmental phenomena, such as the developmental onset of words and phrases, rather than on the basis of true research on developmental pathways to social disabilities. This approach fails to capture important and more subtle developmental phenomena that might earmark true differences between social developmental phenotypes.

Additionally, information regarding early development is often collected retrospectively. This can

have several effects. First, minor abnormalities or specific dates of milestones may not be remembered, particularly if evaluation is made in later childhood or early adulthood. Secondly, minor abnormalities, otherwise a normal part of development, may inadvertently be perceived as significant and 'inflated' if it is believed that they contribute to the diagnosis. If respondents feel they have to provide specific information for diagnosis, then inaccurate information may be given. Therefore, the reliability of the information may be in doubt. Indeed, the data collected for the Field Trial, in which for each case clinicians were asked to provide information about early development in several different ways, and resulted in data that were often conflicting, supports the poor reliability of early developmental information when it is obtained retrospectively. Moreover, too much significance may have been given to Asperger's assertion that the early histories appear to be normal. Whilst Asperger did suggest that the early histories of the cases he described were normal, subsequent analysis did find that 25% of patients he saw and diagnosed with 'autistic psychopathy' had evidence of delay in language and/or cognitive development (Hippler & Klicpera, 2003). Other studies too have found that significant numbers of people with clinical diagnoses of AS have evidence of language/cognitive abnormalities during the first three years (Eisenmajer et al., 1996; Mayes, Calhoun, & Crites, 2001; Miller & Ozonoff, 1997).

Even if there is little sense differentiating groups according to onset, to what extent is the communication domain a useful group differentiating factor? Presently, autism is differentiated from AS on the basis of the presence of specific communication impairments in autism. What is not acknowledged is the fact that impairments of social communication were also described by Asperger, and have been recognized subsequently by all clinicians who have attempted to define the core features of AS. For example, Wing (1981) described the speech as 'pedantic' and 'lengthy', Gillberg (1989) described the 'superficially perfect' expressive language, but which is 'formal' and 'pedantic' and having 'odd prosody' and 'peculiar voice characteristics'. Whilst such impairments of pragmatic language might interfere with the ability to initiate and sustain conversation, and/or result in repetitive patterns of communication (the core criteria for AD), arguably, they are fundamentally different from the type of language and communication impairments described in autism, in which language is delayed, echolalic, idiosyncratic and repetitive. Besides not encapsulating the syndrome completely, omitting these pragmatic communication impairments may cloud the distinction between AS, autism and other disorders of social interaction; for example, schizoid personality disorder, a disorder of social interaction, is characterized by social impairments and rigid patterns of behavior similar to AD and AS, but does not have the pattern of communication difficulties described by Asperger (see Wolff, 2000, and therein). A further issue is that implicit in having the domains of social interaction and behavior defined identically in AD and AS is that the two are related. As discussed above, the relationship between these two is presently poorly understood, and this overlap in phenotypic definition simply confounds any investigation that attempts to validate AS externally.

This distinction between AD and AS notwithstanding, a further issue that needs to be considered is whether, if the two subtypes can be reasonably differentiated phenotypically, they can also be described as forming orthogonal dimensions of social disability. The suggestion would be that AD represents one disorder of social interaction, and is characterized by one pattern of communication deficits and behaviors, and AS is a different disorder of social interaction with its unique pattern of communication and behaviors. The alternative hypothesis would be that the two overlap and form a spectrum, with any differences in behavior or communication a result of cognitive or other differences. Clearly, this idea of different social disabilities requires further investigation, but the results from the present analysis, whereby the majority of cases belonged to either of several groups, suggest that this may be the case.

The recommendation to include AS as a formal diagnostic category was controversial and this controversy was reflected in both the final definition and text adopted by DSM-IVTR. The latter has been very significantly revised in the text revision of DSM-IVTR although criteria remain unchanged. What is needed is an acknowledgement that the onset criteria, as currently defined, are unreliable differentiators of autism and AS, and do not reflect a differentiation between autism and AS on the basis of research on developmental pathways, but, rather, have been set arbitrarily. What is also needed is the recognition of the communication abnormalities of AS. Any future reworking of the diagnostic criteria for AS needs to acknowledge these points and provide more clinical detailed description to allow diagnosis to be made

more reliably. If this was the case, the precedence rule may become redundant. Indeed the original recommendation, not adopted in DSM-IV, had been to make the two disorders mutually exclusive rather than to make one have precedence over the other.

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