Asperger syndrome and high-functioning autism*

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This paper reviews the history of Asperger syndrome (AS) and high-functioning autism (HFA), current diagnostic concepts and criteria, some controversial diagnostic issues, epidemiology, background factors, outcome, and intervention guidelines.

HISTORY OF THE CONCEPTS

Kretschmer's 'schizoid character', 'schizothymia in average people' and 'detached idealism' (Kretschmer, 1924) share many features with AS. Wolff has drawn attention to the report on six boys with 'schizoid personality disorder' by a Russian neuropsychiatrist (Ssucharewa, 1926; Wolff, 1996), who felt the condition bore considerable resemblance to Kretschmer's schizoid variants. These boys appear to have met current criteria for AS (Gillberg, 1991). Wolff's own clinical research has concerned a group of 'schizoid' children, many of whom meet criteria for AS (Wolff & McGuire, 1995).

The Viennese paediatrician Hans Asperger – who, in the early 1940s, was not aware of the Baltimore psychiatrist Leo Kanner's description of children with 'autistic disturbances of affective contact' (Kanner, 1943) – believed that he described a new psychiatric disorder in children in his first paper referring to 'autistic psychopathy' (Asperger, 1944). In the literature published before the 1980s, apart from the writings by Asperger himself (e.g. Asperger, 1952, 1961), I have found four papers referring to 'autistic psychopathy' (van Krevelen & Kuipers, 1962; van Krevelen, 1971; Wurst, 1974; Dauner & Martin, 1978). Robinson and Vitale (see Kanner, 1954) from the USA described three children with circumscribed interest patterns, and at least one, possibly all three, of these met current criteria for AS. Kanner (1954) referred to work (published in German) by Schneerson in Tel-Aviv on 64 8- to 13-year-old children (48 boys and 16 girls) showing similar traits. Kanner did not mention Asperger's work, while Asperger was struck by the similarity of his own and Kanner's cases (Asperger, 1950), even though he later stated that he was referring to a 'basically different type' (Asperger, 1979).

No papers published before 1980 referred expressly to HFA, although, indirectly, many cases with HFA have been implicated in the large body of general autism research. According to my review (aided by S. M. Levy) DeMyer *et al* (1981) was first to use the HFA label. Elisabeth Newson has long been referring to 'more able autistic people' (e.g. Hewett *et al*, 1970).

After Wing coined the term 'Asperger's syndrome', interest in this field has grown (Wing, 1981*a*). More than 160 papers and one book (Frith, 1991) have appeared in the English language explicitly dealing with AS or HFA. The bulk of the papers originated in the UK, Canada and Scandinavia. Recently, the concept of AS has become endorsed in Australia and the USA as well. HFA and 'pervasive developmental disorder not otherwise specified' (PDDNOS) (American Psychiatric Association, 1987) appear to have been more generally adopted in the USA. However, AS has now become part of the international diagnostic nomenclature (World Health Organization, 1992, 1993; American Psychiatric Association, 1994), and it is expected that as operationalised diagnostic criteria (Gillberg & Gillberg, 1989; Gillberg, 1991; World Health Organization, 1993) have become available, the literature will expand at an even greater rate.

Unfortunately, PDDNOS, Rett syndrome and Asperger syndrome are all classified as 299.80 in the DSM–IV (American Psychiatric Association, 1994), and this may contribute to diagnostic confusion.

DIAGNOSTIC DEFINITIONS AND ISSUES

At present, it is not clear that AS and HFA represent distinct conditions. Much of the published work employs diagnostic criteria (World Health Organization, 1993; American Psychiatric Association, 1994) that would make dual diagnosis impossible. My own position is different and includes the possibility that in some individuals it might be appropriate to diagnose autism at one point in time and AS at another.

There are currently no explicit diagnostic guidelines for HFA. It would seem appropriate to diagnose this condition in cases for which autistic disorder (American Psychiatric Association, 1994)/childhood autism (World Health Organization, 1993) criteria apply and the total IQ is above 65–70. This has been the most commonly used definition. Alternatively, a case could be made for a diagnosis of HFA in individuals who have 'mild' autism (e.g. according to the Childhood Autism Rating Scale (Schopler *et al*, 1988)). Whether or not such cases are identical to or essentially different from those diagnosed with AS is open to speculation.

Some studies (Gillberg, 1989; Szatmari *et al*, 1989; Ozonoff *et al*, 1994; Ramberg *et al*, 1996; Ehlers *et al*, 1996) suggest that it is IQ (perhaps specifically verbal IQ) that distinguishes AS (higher verbal and full-scale IQ) from HFA (lower full-scale IQ). It is possible that a positive family history is more typical of AS and less obvious in HFA (Gillberg, 1989).

It is possible that, with IQ controlled, motor clumsiness may be typical of AS (Gillberg, 1989), although this is not generally agreed (Ghaziuddin *et al*, 1992, 1993; Manjiviona & Prior, 1995). However, in the two studies that have suggested no difference, full-scale or performance IQ has been higher in the AS than in the HFA group, making it difficult to draw conclusions. It is conceivable that individuals with AS may be more 'high functioning' than those with HFA, and that abnormal *social* motor control and apraxia may be more evident, because of the wider range of abilities shown by people with AS than by those with HFA.

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SOME CONTROVERSIES IN DIAGNOSIS

The most controversial issues in the diagnosis of AS versus HFA appear to be whether or not:

(a) motor skills should be regarded as a differentiating feature;

(b) AS or HFA could be associated with cognitive disability;

(c) language is impaired in HFA but spared, or even hyperfunctioning, in AS;

(d) a diagnosis of HFA and of AS can be made in the same individual at different stages of development; and

(e) HFA and AS refer to the same or distinct groups of individuals.

The exploration of issue (a) is rendered difficult by the circularity of the arguments used to propose one or the other set of diagnostic criteria. Gillberg's (1991) criteria require the presence of motor control problems for a diagnosis of AS but not for HFA. Thus, a diagnosis of AS would entail more motor control problems than a diagnosis of HFA (which would not require such problems). One study which tried critically to evaluate this problem showed more motor control problems in AS than in HFA (Gillberg, 1989), even in cases for which no prerequisite for motor control problems had been made for the original diagnosis of AS. Motor control problems were an important constituent of Asperger's own gestalt of the syndrome (Asperger, 1944).

The issue (b) pertaining to cognitive dysfunction is contentious. The World Health Organization (1993) research criteria for AS are not compatible with general retardation, and Asperger's own view (most of the time) was similar. Asperger's cases were not always free of major cognitive deficits (case Ernst K in his original paper). Most studies of AS include a few cases with considerably or slightly below average IQ. In clinical practice, a diagnosis of AS is rarely made unless IQ is above 70.

Perhaps the most difficult diagnostic issue (c) relates to early language development. The ICD–10 (World Health Organization, 1993) and the DSM–IV (American Psychiatric Association, 1994) both posit that there be no signs of early language retardation or abnormality and say nothing about later speech or language problems. Gillberg (1991) proposed that some peculiarity of speech and language had to be present. This is in line with Asperger's own view (both Ernst K and Hellmuth L of the original paper had some speech and language delay). Forty-four per cent of Wolff's 32 high-functioning schizoid children (most of whom appear to meet ICD–10 criteria for AS) had "abnormalities of language development" (Wolff, 1991*b*). The ICD–10 criteria for normal early speech–language development are difficult to ascertain in practice (Ehlers & Gillberg, 1993). Most cases of AS are brought to the attention of specialists only at age seven years or later (Gillberg, 1989), when parents may have difficulty remembering whether their child's language development showed signs of abnormality in the early years (Hart *et al*, 1978).

In respect of the issue (d) of 'dual diagnosis' and (e) of the distinction of one syndrome from the other, there are important caveats. In clinical studies it would not make much sense if HFA and AS were compared on a group-wise basis and both groups contained individuals that also had the other diagnosis. This would argue in favour of using diagnostic criteria that would be mutually exclusive for the two diagnoses. This is the position of ICD–10 and DSM–IV. However, we do not yet know which (if any) are the distinguishing features. Exclusion criteria are therefore arbitrary and tend to reinforce the notion of a difference (e.g. if AS is diagnosed only in cases without language problems and HFA only in the presence of such problems, comparative studies would find fewer language problems in AS than in HFA). Large-scale population studies of all aspects of the various problems encountered on the autism spectrum are required to determine to what extent the two concepts are different.

It is of some interest that none of Asperger's own cases meet the DSM–IV criteria for Asperger syndrome (Miller & Ozonoff, 1997). There may be a need for (slightly) different criteria and guidelines for clinical practice versus research. In clinical work, one of the most important points is to arrive at the diagnosis which, at the time and in the given demographic/cultural circumstances, will be of most help to the individual and his/her family. This diagnosis has to be rooted in empirically derived knowledge, but should not be used for splitting academic hairs.

DIFFERENTIAL DIAGNOSIS AND COMORBIDITY

The differential diagnosis in HFA/AS involves many named psychiatric disorders listed in DSM–IV and ICD–10. In addition, the boundaries *vis-à-vis* some learning and language disorders are blurred.

Many disorders in the diagnostic manuals appear not to have been discussed across the different task forces for the various groups of diagnoses. For instance, several of the criteria for obsessive–compulsive personality disorder (OCPD) overlap (albeit slightly differently worded) with those of autistic disorder and AS. This is to be expected, given that many psychiatric disorders lead to restriction of social functioning and this is a symptom included in both OCPD and AS. However, few clinicians (and researchers) appear to be aware that an individual with AS often meets full or almost full criteria for OCPD. Similar cases could be made for schizotypal and paranoid personality disorders. Sometimes this is not just a question of a lack of semantic distinction, but an issue of true comorbidity (Caron & Rutter, 1991).

In a population study of AS, Tourette syndrome was found in one of five school-age children with AS. Tics were demonstrated in another three (Ehlers & Gillberg, 1993). In a population study of anorexia nervosa (Råstam *et al*, 1989), AS was diagnosed in six of 51 cases in early adulthood (Gillberg *et al*, 1994*a*).

Children with deficits in attention, motor control and perception (children who also meet criteria for attention-deficit hyperactivity disorder) occasionally meet full criteria for AS. In one study, three of 14 children with a severe problem of this kind also met full criteria for AS (Gillberg & Gillberg, 1989) and a further five had some autistic traits (Gillberg, 1983). Many children with AS are overactive and aggressive (Eaves *et al*, 1994). The diagnostic overlap is illustrated in Figure 1. In most of these conditions it is likely that some kind of common brain dysfunction/functional variability constitutes the core, rather than the clinical syndromes being distinctive and clearly separable in all cases. One problem common to the children in which there is considerable overlap of diagnoses is in the field of empathy. Gillberg (1992) launched the concept of 'disorders of empathy' to comprise all the groups of clinical syndromes in which reciprocal social interaction in real-life settings is severely impaired.

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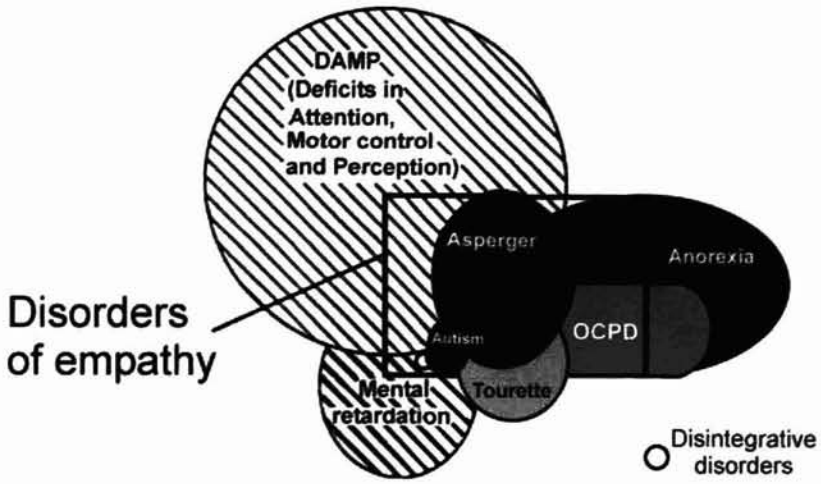


Fig. 1 Differential diagnosis in Asperger syndrome. Overlap with other clinical syndromes. OCPD, obsessive–compulsive personality disorder.

Elective mutism has been described in the near relatives of children with autism and AS (Gillberg & Forsell, 1984; Gillberg, 1989), in girls with AS (Kopp & Gillberg, 1996) and in several 'schizoid' (strongly resembling AS) children examined by Wolff (e.g. Wolff & McGuire, 1995). The possibility of diagnostic overlap of AS/HFA with childhood-onset schizophrenia has also been mentioned (Gordon *et al*, 1994).

Shea & Mesibov (1985) highlighted the convergence of HFA, AS and some forms of learning disability. Children with deficits in attention, motor control and perception usually have mild–moderate learning problems. Their neuropsychological test profile often infers a 'right-hemisphere dysfunction syndrome' (Rourke, 1988; Ellis *et al*, 1994) or a 'non-verbal learning disability'. It is not clear to what extent such a syndrome is equivalent to, overlapping with or different from AS/HFA. The same holds for so-called semantic–pragmatic disorders (Bishop, 1989; Lister-Brook & Bowler, 1992).

Some young adolescents with AS/HFA may develop catatonic features, raising the possibility that in some instances of catatonia in adult psychiatry, AS/HFA may be an antecedent or comorbid problem.

The rate of AS (using Gillberg & Gillberg (1989) criteria) in a male population at a maximum security hospital was 1.5%. The authors proposed that this high prevalence suggested that individuals in forensic psychiatric services should be examined with a view to diagnosing AS/HFA more often than is currently the case (Scragg & Shah, 1994).

EPIDEMIOLOGY

Autism occurs in 7–16 per 10 000 children (Wing, 1993; Gillberg, 1995; Baron-Cohen, 1995). HFA constitutes only a fraction (11–34%) of such cases. Thus, the prevalence is likely to be less than 0.5 in 1000 children in the general population.

The rate of AS, diagnosed on the basis of Gillberg & Gillberg (1989) or ICD–10 criteria, appears to be much higher. According to the only published population study, it is at least 3.6, possibly 7.1 in 1000 children aged 7–16 years (Ehlers & Gillberg, 1993). Wolff *et al* (1991) found that 3–5% of a general child psychiatric clientele were 'schizoid' (often equivalent with AS).

If these figures can be replicated by others, and if AS is accepted as an autism spectrum disorder, the notion that 75% of all children with autism (broadly defined) have concomitant mental retardation would have to be revised. The rate of associated mental retardation in autism spectrum disorders (comprising autistic disorder and AS) would drop to about 15%.

Gender ratios

The gender ratio is skewed both in HFA and AS. In reviews of 16 population studies of autism Wing (1993) and Gillberg (1995) found that the male : female ratio was closer to 2–3:1 than the often-quoted 4–5:1 ratio. However, in HFA the ratio is probably higher (Wing & Gould, 1979). The male : female ratio in autism tends to go down with decreasing IQ (Wing, 1981*b*). In AS clinical studies have suggested a male : female ratio of 10–15:1 (Wing, 1981*a*; Gillberg, 1989), but a population study suggests that it may be in the order of 4:1 instead (Ehlers & Gillberg, 1993).

The underlying reasons for skewed gender ratios in HFA and AS have been proposed to be (a) genetic, (b) brain damage (occurring at about the same rate in boys and girls) in the more severely cognitively disabled accounting for lower male:female ratios (Wing, 1981*b*) in this subgroup, and (c) differences in overall cognitive and social skills in males and females leading to males more often having a style characterised by poorer social skills, so that in the presence of empathy problems they would be more likely to be recognised in males because of their relative lack of 'social camouflaging' abilities (Gillberg, 1992).

BACKGROUND FACTORS

It is not clear what causes HFA or AS. There is general agreement that brain dysfunction is at the root of autism. There appears to be consensus that AS also is caused by brain variation/dysfunction (Frith, 1991).

Cognitive peculiarities

HFA is associated with a particular pattern of peaks and troughs at neuropsychological testing (Lockyer & Rutter, 1970; Rumsey & Hamburger, 1988; Frith, 1989; Happé, 1994; Ehlers *et al*, 1996). Five studies have detailed systematic data on IQ in individuals with AS (Wurst, 1974; Gillberg, 1989; Szatmari *et al*, 1990; Fine *et al*, 1991; Ehlers *et al*, 1996). Overall IQ tends to be higher than in HFA, even when the requirement for inclusion in the study has been an IQ of 70 and above (Ehlers *et al*, 1996). Verbal IQ is often higher than performance IQ in cases with AS, and the reverse is common in cases with HFA. This might be an artefact introduced by the diagnostic criteria applied. 'Verbal' is not necessarily equivalent with 'speech and language'; speech and language problems, particularly in domains of pragmatics conversation, may be as prevalent in AS as in HFA, once IQ is controlled for (Ramberg *et al*, 1996).

Mentalising ('theory of mind') deficits are very common in young children with

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HFA (Baron-Cohen *et al*, 1985). Mentalising refers to the ability, present in normal children of pre-school age, to attribute mental states (thoughts, beliefs, feelings) to others and to oneself. Young children with AS also may have problems in this domain (Frith, 1991; Happé, 1994). As they grow older, they tend to pass theory of mind tasks on formal testing (Prior *et al*, 1990; Ozonoff *et al*, 1991; Bowler, 1992; Happé *et al*, 1996) but not necessarily in real-life settings.

Frontal lobe dysfunction in AS and HFA has been inferred from studies demonstrating executive function deficits (Rumsey, 1985; Rumsey & Hamburger, 1988; Prior *et al*, 1990; Ozonoff *et al*, 1991). McEvoy *et al* (1993) reported a significant relationship between executive function and social communication skills in a group of pre-school children with HFA.

The ability to use cohesive links to create a reciprocal conversation was impaired in HFA and (to a lesser degree) in AS (Fine *et al*, 1991). In a small study of three adults (male), the inner experiences of people with AS were characterised by visual images in contrast to more variable experiences in people without the syndrome (Hurlburt *et al*, 1994). In another study, using Asch's line judgement, AS subjects were more likely to adopt a consistently conforming or non-conforming strategy in an experimental social situation (Bowler & Worley, 1994).

AS and HFA in children tend to be associated with facial perceptual as well as other visual perceptual problems (Davies *et al*, 1994). Failure to make use of configural information in faces may lead to reduced facial perceptual capacity. The results may be taken to support Frith's (1989) notion – based on hypotheses previously formulated by Hermelin & O'Connor (1970) – of a crucial characteristic of autism, viz. the lack of a drive for central coherence.

Developmental/congenital prosopagnosia (the inability to recognise faces) has been proposed as one possible underlying neuropsychological core deficit in certain cases of AS (Kracke, 1994). Gaze avoidance in AS may not be absolute, but may rather be interpreted as a lack of *expected* gaze (such as when another person is talking) (Tantam *et al*, 1993). A life-long absence of gaze response to social cues (including speech) might explain a number of features of the syndrome including decreased joint attention and response to affects of others, and poor discrimination of facial expression.

Impairment of the ability to access stored knowledge has been suggested to be one of the basic deficits in autism, and this, rather than a 'representational' (or theory of mind) impairment, could account for the problems encountered in relatively able people with autism (Lewis & Boucher, 1995).

In summary, at least four different cognitive models have been proposed to be the brain–behaviour mediator in HFA and AS, viz. mentalising, central coherence, executive function deficits and generativity. All four capacities may be impaired, at least in young children with HFA and AS. The dysfunctions are not mutually exclusive and, depending on the degree of impairment in each domain, the clinical profile may vary from one case to another. It is possible that the four domains overlap considerably and that they do not all represent distinct and clearly separable neural functional circuitries.

Brain dysfunction and associated medical disorders

Wing (1981*a*) reported that 47% of her AS cases had indications of severe pre- or perinatal distress. Gillberg (1989) reported 43% of 23 referred AS patients and Rickarby *et al* (1991) found 67% of a referred group of 12 AS patients had some index of major perinatal distress. However, some of these problems could be symptoms of constitutional defects in the foetus rather than markers of brain damage that would lead to AS. In one study (Gillberg *et al*, 1987), 75% of young children and adolescents with HFA (n=17) or AS (n=3) had some index of brain dysfunction and of these 75%, three had an identifiable medical syndrome. Several case reports have documented specific medical disorders in AS/HFA, including tuberous sclerosis (Gillberg *et al*, 1994*b*; Williamson & Bolton, 1995), Marfan-like syndromes (Tantam *et al*, 1990), Kleine–Levin syndrome (Berthier *et al*, 19921), fragile X syndrome (Hagerman, 1989) and other chromosomal syndromes, including 17–19 translocation (Annerén *et al*, 1995).

The rate of brain dysfunction appears to be lower in AS than in HFA, but the scope and meaning of this difference is unclear. At present, almost all studies in the field have been on potentially biased samples, and no general conclusions are warranted.

The rate of epilepsy may be increased compared with individuals without autism. In one study (Gillberg, 1989) the rate was 4%, and in another (Ehlers *et al*, 1996) it was 0% in AS and 10% in HFA.

Laboratory findings

In comparing the computed tomographic brain scans of 18 children with AS and 22 with HFA, Gillberg (1989) found some cerebral atrophy in 17% of the former and 22% of the latter group. Berthier *et al* (1993) found brain abnormalities (cortical, mostly right sided and ventricular enlargement) on magnetic resonance imaging in five of seven young adult males with AS (who had Tourette syndrome in addition to AS). Males with HFA had larger mid-sagittal brain areas than age- and IQ-comparable controls and than males comparable for age and socio-economic status (Piven *et al*, 1992).

A single photon emission computed tomographic study of 12 children with HFA but without epilepsy and 10 children with low-functioning autism and epilepsy showed left and bilateral temporal hypoperfusion in all cases and some frontal hypoperfusion in 36% of the cases (Gillberg *et al*, 1993). In another such study, temporal lobe abnormality was common in the group with autism. However, in that study, only two of 18 individuals appear to have had HFA (Chiron *et al*, 1995)

In one of three studies with positron emission tomography (PET), interactions between frontal/parietal regions and the neostriatum and the thalamus were abnormal (Horwitz *et al*, 1988). The other PET study yielded no significant differences between an HFA and a normal group of adults (Herold *et al*, 1988).

In a controlled collaborative Anglo-Swedish PET study, normal young men showed increased regional blood flow in a specific area of the left frontal lobe in connection with mentalising tasks, whereas young men with AS showed increase in a completely different area of the frontal lobe. This preliminary study suggests that mentalising may be associated with activation of a functional entity in the medial portion of the left frontal lobe and that this entity may be dysfunctional in AS (Happé *et al*, 1996).

Similar unspecific abnormal electroencephalogram, auditory brainstem response and oculomotor findings have

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been reported in HFA and AS as in low-functioning autism (e.g. Gillberg *et al*, 1987; Gillberg, 1989).

The ratio of homovanillic acid (the breakdown product of dopamine) to hydroxymethoxyphenylglycol (the breakdown product of noradrenaline) was increased over control levels and equal to levels in low-functioning autism in one study including a small number of children with HFA (Gillberg *et al*, 1987). In a study of cerebrospinal fluid glial fibrillary acidic protein (GFA-p), a small group of children with AS (n=4) had contents of his glial/synapse marker which were in between those of a normal group (n=10) and an autism group (which included an HFA subsample (n=14) with high GFA-p levels equal to those of the low-functioning cases) (Ahlsén *et al*, 1993). The ganglioside GM-1 was significantly raised in the cerebrospinal fluid of children with autism and autistic-like conditions, particularly in those with HFA and AS (further details available from the author upon request). The findings have been interpreted as possibly reflecting synapse dysfunction in autism spectrum disorders.

Genetic factors

Many cases of autism are caused by genetic factors (Bolton & Rutter, 1990). Twin studies show a striking concordance for autism in monozygotic twins and discordance in dizygotic twins, but it is too early to conclude that almost all autism cases are caused by genetic factors (alone or acting in conjunction with other factors). Findings obtained in twins are not necessarily generalisable to the general population of non-twin autism cases. Also, the twin studies performed have excluded twin pairs in which there was a known medical disorder associated with autism.

It has been hypothesised that genetic factors might be particularly important in the high-functioning cases of autism given that these show less clear-cut associations with indices of brain damage (see above) and the conspicuously high male : female ratio in this subgroup (Wing, 1981*b*). Nevertheless, familial loading was most pronounced in cases with autism and severe mental retardation (and in those autism probands with speech who had the highest frequency of autism symptoms) in one study (Bolton *et al*, 1994). Familial loading was extremely low (2.8% for a broad autism spectrum phenotype) in the HFA subgroup with verbal abilities in the normal (or low-normal) range. However, the sample examined was recruited from a tertiary autism centre, only 29% were non-speaking (compared to about 50% in other samples) and individuals with IQs under 30 appear to have been excluded (such low-IQ cases constitute a considerable proportion in population samples). Thus, no conclusive evidence exists at this stage, and there is a need for more genetic studies on population samples of probands with AS/HFA (and low-functioning autism) and their relatives.

There have been several case reports (Burgoine & Wing, 1983; Bowman, 1988; Gillberg, 1991; Ghaziuddin *et al*, 1993) and at least two formal (clinical, not population-based) studies (De Long & Dwyer, 1988; Gillberg, 1989) that suggest the importance of familial transmission of AS. In Gillberg (1989) there was more 'AS-like' pathology in the close family members of children diagnosed with AS compared with those receiving a diagnosis of HFA. In addition, Wolff's study of schizoid children found a high proportion (more than half) of the parents to be themselves schizoid (Wolff, 1991*a*,*b*). This supports the notion of AS being a predominantly genetic disorder and HFA being either a genetic disorder with a different inheritance pattern or more often caused by brain damage. Other studies suggest that a broader/milder autism phenotype might be present in many first-degree relatives of children with classic (including low-functioning) autism (Folstein & Piven, 1991). Pragmatic language deficits in parents with autism may be another expression of the same genetic liability as for autism in some families (Landa *et al*, 1992). DeLong & Dwyer (1988) and De Long & Nohria (1994) have presented preliminary evidence that children with HFA/AS often have close relatives with bipolar and unipolar affective disorders; De Long proposed that HFA/AS may sometimes represent the earliest, most severe variant of an affective disorder. Gillberg (1985) described the case of an adolescent boy with HFA and bipolar psychosis.

Gillberg (1992) has suggested that empathy might be a normally distributed trait in the general population. Hypothetically, being born to parents functioning at the lower end of the normal distribution of empathy would increase the statistical risk of empathy skills poor enough to warrant a diagnosis of AS. Perhaps with the addition of brain damage, such cases might present as classical autism, or brain damage could act alone to cause AS (mild damage) or HFA (more severe). This general hypothesis is close to the model suggested by Van Krevelen (1971), and by Asperger himself (e.g. Asperger, 1944). Only population studies will allow generalisable conclusions regarding the proportion of all AS cases that have a predominantly genetic background.

Thus, there is widespread agreement that AS is probably a genetic disorder/variant in a majority of all cases. Nevertheless, studies reporting on series of clinical cases (Wing, 1981*a*, Gillberg, 1989, Rickarby *et al*, 1991) have all found a high index of perinatal distress, and one of these studies (Rickarby *et al*, 1991) reported no indication of familial loading. Therefore, any generalised conclusion in this respect would seem premature. The same line of reasoning appears to be valid in HFA.

Social factors

Autism does not appear to be correlated with social class (Schopler *et al*, 1979; Wing, 1980, 1993; Gillberg & Schaumann, 1982; Gillberg, 1991), but it is too early to decide whether this is equally true of HFA/AS as of low-functioning autism. No studies have inferred any particular type of social/family dysfunction in HFA/AS.

Migration from geographically distant cultures has been suggested to be correlated with autism. No studies implicating such migration as a potential contributory factor in the causation of HFA or AS have been published. In the Swedish population study by Gillberg *et al* (1991), out of 35 DSM–III–R autism cases, five were HFA in the sense that they had full-scale IQ over 70. Of these, two had at least one immigrant parent. This figure was similar to that obtained in the remaining group of 30 lower-functioning cases (13/30=43%).

The fact that some parents of children with HFA and AS themselves have autism-associated features begs the question of parenting skills in such individuals. It would not be unreasonable to assume that poor empathy in the parent might contribute to some behavioural/psychological problem in the child, quite apart from any genetic influence. However, one might equally argue that a parent with similar but milder problems would be better able to understand and cope with some of the child's problems because these may be perceived as personality style rather than

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'disorder'. Future studies should seek to explore these issues and try sensibly and sensitively to avoid the mistakes of the past regarding the scape-goating of parents.

WORK-UP

All people with HFA/AS should have a neuropsychological/medical work-up. This will need to be tailored to the age and general functioning of the individual. All children (but not all adolescents and adults) with any of these diagnoses will need: (a) at the very least, testing with a well-validated IQ-test; (b) a medical work-up, comprising a meticulous physical examination (looking for possible signs of neurocutaneous disorders and syndromes associated with particular physical phenotypes), a chromosomal culture and a polymerase chain reaction analysis for the fragile-X gene; and (c) in cases with low IQ or if there is a clinical suspicion for other reasons, a neuroimaging examination of the brain, plus any other laboratory work-up considered to be appropriate (depending on possible additional problems such as epilepsy, motor deficits and metabolic or neurological symptoms).

Probably under 15% of HFA/AS cases have an associated medical condition (e.g. the fragile X syndrome, tuberous sclerosis, neurofibromatosis, hypomelanosis of Ito). In those who do, it is essential to know about it; specific genetic counselling pertains to some of them, certain treatments are more beneficial and outcome varies considerably depending on which specific disorder is involved. There is often no way of diagnosing such disorders without a work-up of the kind outlined.

OUTCOME

The strongest predictor of poor outcome in autism is low IQ. This indicates that higher IQ (roughly to be equated with HFA) would be associated with better outcome. However, in autism outcome studies (reviewed in Gillberg, 1991), absolute numbers of HFA cases have been low in each study, precluding conclusions in respect of the highest functioning group. Furthermore, no study to date has systematically studied outcome in AS, which is probably considerably more common than HFA. The indications regarding outcome gained in studies of autism may not generalise to this population.

AS can be associated with excellent outcome (at least in terms of self-care and academic progress) (e.g. Gillberg, 1991; Sacks, 1995). About half of a total population of AS cases were not referred to doctors or psychologists at school age, even when problems were well recognised by teachers (Ehlers & Gillberg, 1993). Unlike autism, which usually would prompt referral to some kind of specialist before school age, AS may remain undiagnosed well into adulthood. Even then, it is more likely that, if referral is made, the diagnosis will not be AS, but may come out as schizoid or schizotypal personality disorder, type II schizophrenia, atypical depression, paranoid disorder or obsessive–compulsive disorder.

In one population-study that included cases with AS (Hellgren *et al*, 1994), two out of three individuals had severe alcohol problems already at age 16 years.

Rumsey *et al* (1985) described the progress of 10 HFA males, four of whom had been diagnosed by Kanner, from childhood through to a mean age of 26 (range 18–39) years. Social and behavioural impairment remained, but nine had completed high school and two had completed at least one year at a junior college. All had full-scale IQ over 80, but there were persistent deficiencies in comprehension and picture arrangement; pragmatic language dysfunction and speech dysprosody were also common. This study may have been biased in terms of sample selection, and generalised conclusions cannot be drawn regarding outcome in HFA.

The clinical studies of Wing (1981*a*) and Wolff (Wolff & McGuire, 1995) have suggested a high incidence of suicide in AS.

The retrospective studies by Tantam (1991) of an adult psychiatric in-patient clientele of 'eccentrics' and other individuals showing social oddities, indirectly infer that many adult psychiatric patients may have qualified for a diagnosis of AS in childhood and adolescence. Similar conclusions obtained in a Swedish study by Beier (1993).

The studies by Wolff (e.g. Wolff & Chick, 1980; Wolff *et al*, 1991; Wolff, 1995) on schizoid children and Sparrow *et al* (1986) on 'schizoid' and 'atypical' children, several of whom have AS, suggest that the diagnosis often remains stable over time and impairments in socialisation and communication persist over many years. Positive change is not rare, but a proportion develops additional psychiatric symptoms in adolescence and adulthood. This does not mean that the person with AS will become basically very different over time, but rather that similar or identical underlying problems show in different ways and are perceived and diagnosed differently in adulthood than in childhood.

INTERVENTION

There is no cure for most individuals from HFA or AS. In the case of AS in some young children, it is questionable whether a complete cure should be sought. If, as has been implied by Gillberg (1991) and Sacks (1995), some individuals with AS, without extreme personal suffering, go on to become adults who are (albeit in a limited area) creative and contribute to development in their field, it might be unethical to speak of 'treatment'. However, from school age and adolescence, individual and family suffering is often such that some kind of management strategy will need to be considered.

Many of the management guidelines suggested for individuals with autism by Rutter (1985) and Gillberg (1988) apply in HFA and AS as well (see Appendix).

Clomipramine (Gordon *et al*, 1993) was effective in reducing rituals, obsessions and compulsions in a double-blind placebo-controlled study of 24 individuals, most of whom had HFA. Some individuals with AS have severe attentional problems and it is possible that these could benefit from central stimulant treatment (Strayhorn, 1989). Studies examining the effects of the selective serotonin reuptake inhibitors in AS/HFA are currently in progress. Positive results from open studies still await confirmation.

Psychotherapy and pharmacological treatment are usually not indicated in HFA/AS. Occasionally, both can be clinically useful, singly or in combination. All interventions should be managed by clinicians well informed about the nature of the underlying disabilities.

Ozonoff & Miller (1995) tried to teach theory of mind skills to young people with HFA. Interventions were successful in achieving meaningful change on several tests of theory of mind, but no change in social competence was noted by parents or teachers.

Social skills groups (Mesibov, 1984; Marriage *et al*, 1995) may be useful for promoting 'superficial' social skills in HFA/

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AS, but, so far, no systematic evaluation of the effects of such group intervention has been published.

In the past, all too often, only the deficits have been highlighted, leading to a pessimistic view in respect of management and outcome. However, most deficits/symptoms can be turned into assets if thoughtfully reconsidered. For instance, the 'over-focus' on a narrow interest might be viewed as an exceptional ability to concentrate, if only the right settings and tasks are provided. The inability to relate normally in a social setting could be seen as an ability to shut out irrelevant social stimuli and might provide the framework for an unusual capacity for hard, rigorous work. Twachtman (1998) and Gray (1996) have described helpful ways of identifying areas of interest and good skills that can be used for teaching other skills. Gray's approach of using 'social stores' to identify problem behaviours and teaching coping strategies could hold particular promise here.

Nevertheless, in some respects, the deficits need to be seen for what they are: disabling problems in the domains of social interaction, communication and behavioural adjustment. Thus, for instance, an extremely bright young man with AS might not be able to head a company or a department, even if his overall IQ would seem to predict a very bright future. And a young woman with HFA/AS with very good artistic skills but an overall IQ of about 90 might not be expected to cope with the simplest social interactions and may have to be accommodated in sheltered housing and vocation in adult life.

CLINICAL IMPLICATIONS

- Asperger syndrome and high-functioning autism are much more common than previously believed.
- Asperger syndrome is sometimes mistaken for other psychiatric disorders, such as schizophrenia, paranoia and personality disorders.
- There is a great need for clinical training of psychiatrists (adult psychiatrists in particular) in the diagnosis and management of autism spectrum disorders.

LIMITATIONS

- There is continued controversy as to the diagnostic boundaries of Asperger syndrome and it is unclear whether it is distinct from or equivalent to high-functioning autism.
- Most of the published work has referred to potentially biased populations and small proband cohorts, rendering generalised conclusions impossible.
- Not much is known about outcome and appropriate treatment/management in high-functioning autism and Asperger syndrome.

CONCLUSIONS

HFA occurs in fewer than 1 in 2000 children. AS may be 5–10 times more common. About one per cent of all children raise suspicion of suffering from autism or autism spectrum disorder (Wing, 1966). All such disorders affect boys more frequently than girls. It is unclear whether autism spectrum disorders represent separate, overlapping, or identical conditions. Research into the epidemiology, differential diagnosis, neuropsychology, neurobiology, treatment and outcome of HFA/AS is needed. AS has a better outcome than lower-functioning autism. However, some individuals with AS have persistent problems and consult adult psychiatrists for them. Adult psychiatrists need to be trained so that they will be able to recognise HFA and AS in patients currently receiving a plethora of diagnoses, ranging from atypical schizophrenia, paranoid disorder and depression to schizoid or schizotypal personality disorder.

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APPENDIX

Management guidelines for high-functioning autism (HFA) and Asperger syndrome (AS).

(a) The first evaluation: appropriate assessment and correct diagnosis essential for optimal understanding and service

(b) Neuropsychiatric work-up: psychological evaluation including IQ, psychiatric evaluation for comorbidity, medical and laboratory work-up, for possible associated medical conditions (fragile X, tuberous sclerosis, hypomelanosis of Ito etc.)

(c) Information to and education of all family members (including siblings and grand-parents)

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on helpful coping strategies (educational and behavioural) plus involvement with support groups (if possible)

(d) Work with the family

(e) Practical and financial support for the family (including respite care)

(f) Education for the patient (including 'theory of mind skills training', special education classrooms for some individuals, and 'HFA/AS consultants' in normal classrooms for others)

(g) Physical education (focusing on social motor skills and 'general exercise')

(h) Social skills groups for some adolescents and adults

(i) Pharmacotherapy in a minority

(j) Psychotherapy in a minority (should only be provided by a therapist well-informed about HFA/AS)

(k) Behaviour modification for particular problems

(l) Long-term perspective

(m) Integration of approaches: the developmental psychiatry team

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