

Asperger's disorder (i)

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WHAT IS ASPERGER'S DISORDER?

One year after Kanner's (1943) original paper on autism, Hans Asperger published a paper in 1944 that formed the basis of what was to become known as Asperger's disorder. Both Kanner and Asperger trained in medicine in Vienna, but, unlike Kanner, who moved to the USA, Asperger remained working in Europe.

Asperger and Kanner were apparently unaware of each other's work, probably because of the Second World War. Asperger's paper remained relatively unknown as it was published in German and was not widely available in translation. The paper described a group of children and adolescents who had deficits in communication and social skills, had obsessional interests and behaviour, disliked change and had a dependence on rituals and routines. In addition, many were physically clumsy. Unlike the children described by Kanner, the children in Asperger's paper generally had no significant delays in early cognitive or language development. Asperger described this condition as 'autistic psychopathy'.

There has been increasing interest in Hans Asperger and his syndrome over the past 20 years. In the early 1990s, Asperger's paper was translated by Frith and became more widely available (Frith, 1991). Since that time Asperger's disorder has been more frequently used to describe a group of children who present with developmental deficits in social skills and behaviour but are difficult to classify.

For the past decade or so there has been a continuing debate as to whether or not Asperger's disorder is a type of autism or whether it constitutes a separate entity. Many publications have tried to delineate the boundaries, if any, between autism and Asperger's disorder. Despite the differences that can

be seen when looking at the original cases described by both Kanner and Asperger, there is continuing confusion over the diagnostic criteria for Asperger's disorder, particularly as subsequent accounts and case studies have not necessarily adhered to the criteria suggested by Asperger himself. The principal areas of inconsistency relate to early development in the areas of cognition, motor skills and language.

In DSM-IV and the ICD-10 there have been attempts to introduce a consistent international approach to diagnosis and to specify that the key differentiation is that persons with Asperger's disorder do not have delayed language development, which is a characteristic of autism. Persons with Asperger's disorder have overall normal intellectual ability. Approximately 20% of persons with autism also have IQ in the normal range and are referred to as high functioning.

HOW COMMON IS ASPERGER'S DISORDER?

Asperger's disorder is thought to be more common than autism. Ehlers and Gillberg (1993) studied a population of Swedish children and suggested an incidence of 36 per 10,000, compared with an incidence of about 10 per 10,000 for autism. Because of the lack of diagnostic clarity, estimates of prevalence should be treated circumspectly. It is very likely that mild degrees of Asperger's disorder may not cause sufficient social, emotional or mental health problems for recognition to be necessary or even desirable, except perhaps for epidemiological purposes. One of the most reliable current estimates of the prevalence of people with Asperger's disorder who have needs for a service is that of Fombonne (1999), who concluded that 20 in 10,000 of the general population met accepted criteria for the disorder. The prevalence might be higher in special populations, for example patients in a high-security hospital. One estimate puts the prevalence in one special hospital at 150 per 10,000.

Some small-scale epidemiological studies suggest that the ratio of males to females with the disorder is about 10:1.

Both prevalence and gender distribution figures require confirmation by larger epidemiological studies and at this stage are only rough estimates.

AGE OF ONSET

Asperger's disorder tends to be diagnosed later than autism in young children. Neither ICD-10 nor DSM-IV stipulates the criteria for age of onset as they

do for autism. However, in his original paper, Asperger described children as having difficulties by the age of two years.

Parents of young children with autism often recognise problems with behaviour and, in particular, language development by about 18 months to two years of age. Because children with Asperger's disorder do not have delayed early language or problems with cognitive development, there are few early signs that all is not well. It is more usual for parents to become concerned about their child's emerging unusual or odd behaviour and social development, but these tend to be identified later, usually from about three to four years of age.

Asperger's disorder may not be diagnosed until the child has attended pre-school or some other early childhood setting such as a crèche. This is probably because the child's social and behavioural problems become more noticeable when the child is seen with peers in a more structured social setting, where there are more demands for social interaction.

DIAGNOSIS

In the interest of avoiding confusion for the person suspected of having Asperger's disorder, the family, clinicians and researchers, it is advocated that the DSM-IV or ICD-10 criteria be applied. One practical answer to the lack of clarity about diagnosis is to break diagnosis down into three steps:

- Does the person have a pervasive developmental disorder?
- Is there a history of delayed language development?
- Is the person of overall normal intellectual ability (i.e. is he or she high functioning)?

Box 2.1. *The triad of social impairments*

Absence or impairment of:

1. comprehension and use of communication, both verbal and non-verbal
2. two-way social interaction
3. true, flexible, imaginative activities, with the substitution of a narrow range of repetitive, stereotyped pursuits

Adapted from Wing and Gould (1979).

This approach has some advantages. It may be more reliable to diagnose the presence of a pervasive developmental disorder than to make a diagnosis of a specific pervasive developmental disorder. Many professionals use Wing and Gould's (1979) triad of social impairments (see Box 2.1) as a guide to the presence of a pervasive developmental disorder and, indeed, elements of the triad are apparent in both the ICD-10 and the DSM-IV criteria.

Another advantage of this pragmatic approach is that it is needs based. More able people with either high-functioning autism or Asperger's disorder do need very different services than less-able people. They are much more self-reflective and are therefore very conscious of their environment. More able adolescents and adults with high-functioning autism or Asperger's disorder may have been sensitised to humiliation and may consequently have become very sensitive to status. They may therefore be very intolerant of receiving help in a mixed group containing people who are much more handicapped than themselves. Finally, more able people with autism or Asperger's disorder may be capable of considerable or even complete independence and autonomy, and the services that they receive should reflect this.

The presentation and skills of a person with a pervasive developmental disorder may change over time and a less able child may grow into a more able adult. Children with autism and normal intellectual ability will probably develop into adults with adequate language ability, although they are still likely to have problems with the social and conversational use of language. In these cases, the diagnosis of high-functioning autism would continue to apply. Although adults diagnosed with high-functioning autism and those with Asperger's disorder are

Box 2.2. Multi-level approach to assessment

Level 1. Examine the client and obtain systematic developmental information. What developmental disorders are present?

Level 2. Talk to the client and a key informant. How have the client's developmental difficulties been affected by the client's emotional reactions to them? How have they been influenced by other factors, such as age, intelligence, anxiety levels and comorbidity?

Level 3. Talk to the client, talk to a family member and consider other professional reports. How has the client's reactions to the disabilities been influenced by other people's reactions to them? Has the client been victimised? What have been the family's expectations of, and reactions to, the client?

likely to have a number of features in common, their differing developmental pathways, particularly with respect to language development, produce neuro-cognitive and behavioural differences.

The adjustment of a person with a pervasive developmental disorder is the end result of the interaction of various neurocognitive disabilities, that person's way of coping with them, and the impact of other people's reactions to them. Applying this 'functional' diagnostic approach leads to a multi-level assessment (see Box 2.2). The profile of specific disabilities so obtained will distinguish Asperger's disorder from autism, but it will also be found that people will differ slightly and will therefore have their own personality and profile of strengths and weaknesses.

WHAT CAUSES ASPERGER'S DISORDER?

There is widespread agreement that genetic factors predominate as the primary cause of Asperger's disorder. Asperger himself noted that in all cases where he studied the family closely, similar traits were found to some degree in parents and other family members. Later studies have found similar autistic traits in the relatives of young people with Asperger's disorder.

SOME EXAMPLES OF HOW ASPERGER'S DISORDER AFFECTS CHILDREN

- Acquisition of language follows a normal or even accelerated pattern, but content of speech is abnormal – it is pedantic and may centre on one or two favoured topics.
- There is little facial expression, vocal intonation may be monotonous and tone may be inappropriate.
- There is impairment of two-way social interaction, including an inability to understand the rules governing social behaviour. The person may be easily led.
- There are problems with social comprehension despite superior verbal skills.
- The person tends to be very rigid and to prefer structure.
- The person has well-developed verbal memory skills, absorbs facts easily, and will typically have a good level of performance at maths and science.
- The person is highly anxious and is likely to dislike any form of criticism or imperfection.
- Most attend mainstream schools and are often victims of teasing, which causes withdrawal into isolated activities.
- The person will be seen to be 'odd' or 'eccentric'.

ARE THERE ANY DIFFERENCES BETWEEN AUTISM AND ASPERGER'S DISORDER?

The simple answer to this question is yes. The recent debate as to whether or not the two disorders differ is clouded because clinicians have not used a consistent set of diagnostic criteria to characterise their subject populations when exploring the differences between Asperger's disorder and autism. This variability of diagnostic assignment has led to a situation where studies examining the validity of Asperger's disorder as a separate disorder, particularly in contrast to high-functioning autism, cannot be easily compared or interpreted. This problem can be addressed by the use of the ICD-10 and DSM-IV definitions and diagnostic criteria.

For example, a recent study of high-functioning autism and Asperger's disorder strictly defined by DSM-IV criteria found that children and adolescents with Asperger's disorder presented with higher levels of overall psychopathology, were more disruptive, antisocial and anxious, and had more problems with social relationships than the children with high-functioning autism (Tonge *et al.*, 1999). These differences were not due to any age or global IQ differences.

The finding of high levels of anxiety and disruptive behaviour in the Asperger's group has particular clinical relevance, as these psychopathological problems are potentially open to treatment. Anxiety symptoms may be responsive to cognitive-behavioural interventions and psychopharmacological treatment. Disruptive behaviours can be modified by educational, environmental and behavioural modification techniques.

In the absence of identifiable neurological damage, neurobehavioural studies have indicated differences between autism and Asperger's disorder. While there are broad similarities in the clinical (e.g. social dysfunction) and neuropsychological (e.g. visual-perceptual processing anomalies) features associated with high-functioning autism and Asperger's disorder, recent research has identified differences in executive functioning, lateralisation and motor ability, and this supports the notion of a differing neurobiological basis. For example, a recent series of experiments has indicated that executive functioning, in particular inhibitory deficiencies, is quantitatively and qualitatively different in autism and Asperger's disorder. Individuals with autism had difficulty inhibiting cognitive-motor responses at increasing levels of task complexity. In contrast, individuals with Asperger's disorder performed similarly to age- and IQ-matched controls. It was noted that a combination of inhibitory and set-shifting deficits might have accounted for performance deterioration in the autism group. Interestingly, past researchers have emphasised only set-shifting, but not inhibitory, deficiencies in autism. Further, young people with autism were significantly

slower at shifting attention from the local to global features of a numerical configuration than those with Asperger's disorder, who had no such difficulty.

Motor functioning

Clinical observation suggests that motor clumsiness is a feature that may distinguish Asperger's disorder from autism. In reviewing the literature, Ghaziuddin *et al* (1994) found that approximately 50% of publications referred to clumsy, uncoordinated movement patterns in either single case studies or group studies of children with Asperger's disorder. Gillberg (1989) observed that individuals with Asperger's disorder 'appeared to be generally clumsy', had a 'stiff or awkward way of walking (often without arm-swing)' and were 'uncoordinated in posture and gesture'. Recent functional magnetic resonance imaging data have revealed that individuals with autism exhibit less pronounced activation in the primary motor cortex and supplementary motor area during a simple finger-tapping task than individuals with Asperger's disorder, who do not exhibit this decrease in supplementary motor area activity but show a prolonged activation following the movement. If the mechanism for terminating one motor movement before another is initiated is dysfunctional in Asperger's disorder, this could account for the clumsiness.