Asperger’s disorder (ii)

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DIAGNOSIS OF ASPERGER’S DISORDER

It was not until the 1990s that there was a consensual, although tentative, definition of Asperger’s disorder, in the DSM–IV and ICD–10 classifications. It is currently defined as a pervasive developmental disorder separate from autism with a set of specific diagnostic criteria.

The ICD–10 and DSM–IV definitions are largely the same, but the ICD–10 guidelines for research are more detailed. The criteria for social impairments and abnormalities in play and behaviour are the same for autism and Asperger’s disorder. The key feature that differentiates autism from Asperger’s disorder is the absence of language delay in children with Asperger’s disorder. ICD–10 also includes two common characteristics – motor deficits and isolated special skills – but these are not necessary for diagnosis (see Box 3.1).

Asperger’s disorder is classified as a developmental disorder, but it is probably more useful to consider that a person who meets either DSM–IV or ICD–10 criteria for it (I shall consider them to be equivalent for the purposes of this chapter) has one or more developmental disorders (see Box 3.2), since the clinical and experimental picture indicates the presence of multiple cognitive problems. It is particularly useful to consider these developmental disorders individually, as the extent to which they contribute to the clinical picture of a person with Asperger’s disorder varies. Each person with Asperger’s disorder therefore has an almost unique profile of pervasive developmental disorders.
Box 3.1. ICD–10 criteria for Asperger’s syndrome (Asperger’s disorder)

A. There is no clinically significant general delay in spoken or receptive language or cognitive development. Diagnosis requires that single words should have developed by 2 years of age or earlier and that communicative phrases be used by 3 years of age or earlier. Self-help skills, adaptive behaviour, and curiosity about the environment during the first 3 years should be at a level consistent with normal intellectual development. However, motor milestones may be somewhat delayed and motor clumsiness is usual (although not a necessary diagnostic feature). Isolated special skills, often related to abnormal preoccupations, are common, but are not required for diagnosis.

B. There are qualitative abnormalities in reciprocal social interaction (criteria as for autism).

C. The individual exhibits an unusually intense, circumscribed interest or restricted, repetitive, and stereotyped patterns of behaviour, interests and activities (criteria as for autism; however it would be less usual for these to include either motor mannerisms or preoccupations with part-objects or non-functional elements of play materials).

D. The disorder is not attributable to the other varieties of pervasive developmental disorder: simple schizophrenia (F20.6); schizotypal disorder (F21); obsessive–compulsive disorder (F42.–); anankastic personality disorder (F60.5); reactive and disinhibited attachment disorders of childhood (F94.0 and F94.2, respectively).


Box 3.2. Psychodevelopmental disorders relevant to an understanding of Asperger’s disorder

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CLINICAL MANIFESTATIONS OF ASPERGER’S DISORDER

Delayed, relative to overall developmental level, or deviant non-verbal communication is the one essential requirement of a diagnosis of an autistic spectrum disorder in general, and of Asperger’s disorder in particular. Problems in social interaction are inevitable if a person has a non-verbal communication disorder. Acquired non-verbal communication disorder, as may occur in schizophrenia for example, may also lead to problems in social interaction but is easily distinguished from Asperger’s disorder because it develops during adolescence or adulthood.

There may be conspicuous impairment of non-verbal expression, or expression may be relatively spared but there is impairment of non-verbal interpretation.

Impaired non-verbal expression

Anthony was delayed in his use and understanding of speech, in his motor coordination, and in his socialisation. His motor difficulties were first picked up in relation to his writing and he was diagnosed as having dyslexia when he was aged seven. Anthony has always had impairments of empathy, probably related to his difficulty in reading other people’s facial expressions, and even now he may say untoward or insensitive things in social situations. He has also had long-standing difficulties in non-verbal expression. Anthony tends to look at his listener more when he is speaking, often to emphasise points. This is contrary to gaze frequency normally, since it is listeners rather than speakers who look most at the other person. Anthony looks close to the other person’s eyes as often as he looks directly at the other person’s eyes. This too is unusual. Gaze is normally either directed into the other person’s eyes or well away from them.

Anthony does not use other-directed gaze to claim his turn when there is a pause in the conversation, and does not use it to indicate his interest in another person’s speech to him. He laughs a lot, sometimes about things that are not funny, but has a limited range of other facial expressions. Odd ‘manneristic’ expressions cross his face when it is in repose. The lower two-thirds of his face are less mobile than his brow. His parents reported that it was difficult to tell what he was feeling from his facial expression as a child. Anthony shows none of the fleeting expressions that cross a person’s face normally when in conversation. Anthony can, however, mimic facial expressions when he wants to and can show happiness or anger by an effort of will. There are abnormalities in his voice prosody: in the tone of voice, the rhythm, and the stress patterns of his speech. There is often a rising tone, rather than the normal falling tone, at the end of his sentences. His voice prosody when he is imitating another person may, however, be normal. Anthony uses gestures but they are not directed towards a person to whom he is speaking. Sometimes they seem to be facilitating his speech production by marking and reinforcing the rhythm of his speech. Anthony’s posture may sometimes be exaggerated, when he is copying one of his favourite television characters, or neutral. It does not express his feelings.
A study carried out by Tantam et al (1993) on adults indicates that people with Asperger’s disorder do not attend to socially significant cues by looking and that this may be one reason why they look less at a speaker than is normal. Tantam suggested that people with Asperger’s disorder may lack the normal human response to attend by gaze or head turn to social cues, such as smiles, speaking or being looked at. This is not a perceptual failure; people with Asperger’s disorder show the same amplitude of evoked potential as do normal people when presented with a slide of a face whose gaze appears to be on them as the ‘oddball’ in a sequence of slides of the same face with gaze averted.

Whatever the precise developmental lesion, or lesions, non-verbal communication plays such an important part in maternal–infant bonding, in the development of speech and in the shaping of early social learning that a child born with impaired non-verbal communication is likely to have other problems in these areas, even if these are not primarily affected. This early constellation of difficulties constitutes a core syndrome of Asperger’s disorder (see Box 3.3).

### Unusual interests and motor incoordination

Unusual interests and motor incoordination are included in this list because both are, to some degree, social disorders. Interests are normally learned by identification: that is, by patterning one’s own attention on that of other people. Identification therefore depends on intact non-verbal communication, and is impaired in people with Asperger’s disorder. This does not fully explain the stereotyped nature of many of the interests of a person with Asperger’s disorder. Most special interests will have a repetitive, list-making, classifying or collecting element. This is probably a source of comfort to people with Asperger’s disorder, as it is to people without it, but it is still unclear why people with Asperger’s disorder should be so anxious. One possible explanation is that their impaired non-verbal communication results in people with
Asperger’s disorder being socially isolated, even if they have a normal family and normal family relationship. Social isolation, for example in solitary confinement or in socially impoverished institutions, is known to increase anxiety and repetitive behaviour.

Rituals and stereotypies are not resisted by people with Asperger’s disorder, and so are dissimilar to obsessions and compulsions in adults with obsessive-compulsive disorder, although perhaps not children with obsessive-compulsive disorder, who are reported to show less resistance. They are not the result of a lack of inhibition because they can be deferred, although if they are it may be at the cost of increased arousal and more ritualising later.

Interests are complex, of course, and so are the special interests of Asperger’s disorder. They may also be a way of coping with the disorder, as in the person with Asperger’s disorder who was studying non-verbal communication, and they may be emotionally significant. They may function as memorials of happy times in the past, or of especially attractive people or ideas.

Motor performance, too, is a matter of identification. The swagger of the braggadocio, the sway of the sexy young woman, the earnest demeanour of the cleric are all learnt performances. The clumsiness of a person with Asperger’s disorder is evident in the gait, but it is often difficult to be specific about what is evident. Wide-based gaits and other suggestions of neurological disorder do occur, but they are by no means universal. It is usually a peculiarity of gait that is striking – and this of course means a gait that is individualistic.

**Impaired non-verbal interpretation**

Most people with a diagnosis of Asperger’s disorder have an impairment of non-verbal expression (see above) and of non-verbal interpretation. But there are people who seem to have a deficit in non-verbal interpretation alone. Many of them are not diagnosed as having Asperger’s disorder but as having a personality disorder. This is usually because their communication problems are not recognised. Very often the affected person tries hard to conceal the lack of understanding, sometimes preferring to be thought of as someone bad or psychopathic than to be thought of as deficient in any way. This hatred of feeling inadequate is common in people with Asperger’s disorder, perhaps because they have often experienced shame or humiliation.

Impaired non-verbal interpretation is more difficult to assess than impaired non-verbal expression. Indeed, it may be missed altogether. It may cause a lack of empathy and this may be more obvious, although it is often attributed to a person’s bad character than to a psychodevelopmental disorder. In Asperger’s disorder a lack of empathy may coexist with a strict moral code, although this
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is not universal. Moral codes are a product of upbringing and while some families inculcate strict values which a person with Asperger’s disorder will, typically, apply unswervingly, other families give much less clear guidance and the children of these families may have a weaker and less coherent set of rules to live by.

Andrew was born after a long labour but seemed to develop normally and was, if anything, too placid. This gradually changed as he got older. He began to have screaming attacks and temper tantrums, and could not be comforted by cuddling. In fact, he seemed to dislike close physical contact. His parents were sometimes struck by Andrew’s lack of response to other people’s feelings and this often caused trouble if another child came round to play. Andrew would end up in a fight and could be quite spiteful. Eventually, other children stopped coming. Andrew was clumsy, did not learn to ride a bike until he was 10 and had problems with dressing until well after he had started at school. He sometimes spoke in an odd tone of voice and occasionally walked on tiptoe or flapped his arms. His words would sometimes be in the wrong order. He copied what he saw on the television and would use phrases he had picked up from his favourite shows. These included American wrestling. He could pretend to be a wrestler, but only in his own way. He did not share these games with others. He had little sense of socially appropriate behaviour and would sometimes sit on the lavatory with the door open despite the fact that there were people coming and going on the landing. His parents were worried about these behaviours and consulted the health visitor and the local doctor, but were reassured. They accepted this reassurance quite readily as none of these problems seemed individually very serious.

Andrew seemed to settle at school at first, but he had more and more difficulty as he got older. He began to be bullied and also got into trouble for some spiteful attacks on other children. These were always very unexpected. Other people thought him immature but not malicious. His face was unlined and until he was about eight or nine some people even thought he seemed angelic. He became more anxious as he got older, and this was reflected in a restlessness in his movements and his gaze, which detracted from his previously serene manner.

Andrew under-performed at school and attended a special school for a period. Intelligence testing showed that his verbal IQ was lower than average but in the normal range; his performance IQ was, however, mildly subnormal. However, Andrew seemed to have more academic problems than his intelligence would have predicted. He never seemed to be able to prepare homework and he also seemed obtuse in the classroom, to the point where he was often excluded. In retrospect it appeared that Andrew had problems in understanding information written on the blackboard. He could not translate this easily into words. Part of the problem may have been due to a reduced short-term verbal memory store. Andrew also had problems in following instructions or sequences of information, partly because of this limited storage and partly because he would easily be distracted. Other frontal lobe signs became clearer as he got older, when it was apparent that he met criteria for ‘dysexecutive syndrome’ (see below).

Andrew became attached to a gang of boys at about 14 and began to use illicit drugs with them. They encouraged Andrew to become more extreme in his behaviour and put him up to aggressive actions which had him excluded from school on a few occasions. By the time that Andrew had left school he had acquired a record of juvenile offences and had developed a bragging manner which concealed his vulnerability. When he found himself in
a social situation, Andrew would try to dominate it, probably because this gave him the best chance of knowing what was happening. If this failed, Andrew would disrupt the situation by saying or doing something outrageous. His psychiatrist speculated that this was because Andrew found it easy to read outrage in people’s faces but could not read more subtle cues.

The author Doris Lessing has written two short books describing ‘Ben Lovatt’, who has so many similarities to Andrew that I sometimes have called this type of Asperger’s disorder the Fifth Child syndrome, after her first book about Ben.

**Intersubjectivity**

Lessing describes her fictional protagonist often feeling out of his depth and out of touch with what another person wanted. Indeed, she makes him a member of another race of human beings, one that he eventually discovers high in the Andes. This failure to share a common ground of emotional and social understanding is commonly called intersubjectivity and is linked by some to an impaired theory of mind, that is, to an inability to account for how other people, with a different social perspective, would react or what they would believe.

Understanding people who are not speaking one’s native language is a matter of being able to understand their words and to reply in an understandable way. It is a pleasant feeling which can produce a completely spurious, and misleading, sense of closeness. My own clinical experience suggests that intersubjectivity is a similar experience of mutual communication, but in this case the communication is non-verbal.

**Semantic-pragmatic disorder**

The ICD–10 criteria for Asperger’s disorder have language delay as an exclusion criterion, but there are a significant number of more able people with autism whose language development is delayed but then catches up and ceases to become a clinical problem in adulthood, although residual semantic problems may persist.

Although the acquisition of speech probably requires a substrate of non-verbally mediated social interaction, speech activates a capacity for symbolically mediated cognition that becomes an independent second signalling system–language. Some aspects of language use remain non-symbolic and are probably mediated by the non-dominant hemisphere and not the language centres of the dominant hemisphere. These pragmatic aspects of speech are therefore considered in this section.
Most people with Asperger’s disorder have pragmatic abnormalities of speech, even when their language is unimpaired. Speech abnormalities include concrete or idiosyncratic interpretation of words, an interest in the components of words and in made-up words, in over-formal grammar with a lack of idiom, and more obviously sociolinguistic problems, like a paucity of language codes for different situations, and a lack of understanding about polite forms of speech, evasions, concealments and a whole variety of aspects of speech that are often now subsumed under ‘theory of mind’. The term ‘semantic-pragmatic disorder’ is sometimes used for this group of psychodevelopmental language problems.

People with a disorder of non-verbal communication always have semantic-pragmatic problems but not everyone with semantic-pragmatic problems has a disorder of non-verbal communication and merits a diagnosis of Asperger’s disorder.

**Dysexecutive syndrome**

Andrew, in the account given above, was reported to have ‘dysexecutive syndrome’. This has been noted in association with attention deficit hyperactivity disorder, with schizophrenia, during the course of recovery from head injury, with aggression in girls, with autism and with Asperger’s disorder. My clinical experience is that it may, like the other pervasive developmental disorders, exist on its own. Recent neuroimaging studies suggest that anterior cingulate dysfunction is implicated in it, although executive functions have also been attributed to the frontal or prefrontal lobes.

Dysexecutive syndrome may involve the translation of knowledge into practice, holding on to a sequence of instructions, and ‘holding information in mind while suppressing a prepotent response’.

Dysexecutive syndrome has a significant effect on daily living.

Malcolm’s family reported that he had difficulty in picking up conversational cues and would often interrupt conversations. He had difficulty in adjusting his conversation to that of other people and so would turn the conversation back to topics of special interest to him, and showed a lack of empathy. He would become upset if this was pointed out, and seemed to be unaware of his lack of contact with others. Malcolm had a similar lack of self-awareness about his inability to follow instructions or agreements. For example, if he arranged to meet someone he would be in the assigned spot at the assigned time sometimes. But if he saw something he wanted to investigate he would do so, and lose all track of time. Nor would he think to contact the person he was supposed to meet. However, he would come to the meeting place at some much later time and wait for the person to show up there. He could not cook or do his washing unless he was constantly supervised because he would become derailed in these activities, too. For example, if he was cooking something on the
stove and he found that it required carrots, he would go in search of them. But, if he did not find them, he would forget about the cooking and go off to do something else, leaving the pan on the stove to burn.

**Attention disorders**

Attention deficit hyperactivity disorder occurs in people with Asperger’s disorder, perhaps particularly in people with the ‘Fifth Child’ form mentioned previously. It is not clear whether attention deficit hyperactivity disorder is more common in Asperger’s disorder than in the normal population, however. There is an overlap between attention deficit hyperactivity disorder, reduced verbal memory store and dysexecutive syndrome.

Other disorders of attention occur in Asperger’s disorder. Indeed, it has been argued that a joint attention deficit is one of the fundamental difficulties caused by the disorder. Joint attention is a reflection of social understanding. Attention can normally be redirected by other people, either intentionally, for example by using a person’s name, or unintentionally, for example by gaze. People with Asperger’s disorder often do not respond to their name being called, and seem to become absorbed in whatever they are doing, or whatever they are looking at. It is not clear whether this represents a problem in shifting attention or a failure to respond to attempted attention shifts by other people.

**Learning and memory disorders**

Memory disorders in autism are complex. A recent review of memory development in childhood has indicated that short-term, autobiographical memory, episodic memory, and metamemory all need to be considered (Gathercole, 1998).

Reduced working memory is often associated with attention deficit hyperactivity disorder but may be more strongly associated with language disorders such as disconnection dysphasia and dyslexia. Memory disorders have a claim to be pervasive partly because of the impact of reduced working memory, which may adversely affect almost any cognitive activity in which storage is involved.

People with Asperger’s disorder and clumsiness often have difficulty remembering who people are, but will remember events or statements. People with autism who have good geographical knowledge will also remember routes. However, this is not a reflection of the conventional distinction between procedural and declarative memory. People with Asperger’s disorder do not learn by following instructions, but by trial and error or by imitation. This is one
reason why social skills training has been so disappointing and why role-playing may be a more successful approach. Many families say that their child with Asperger’s disorder is the one who best understands how to programme the video, but that this has been achieved without ever looking at the instruction manual. The person with Asperger’s disorder remembers a procedure, but it is a mechanical one and does not involve much social knowledge.

People with Asperger’s disorder are impaired in acquiring and retaining social ‘knowledge’, perhaps because social knowledge is actually based on identification. Note that identification is not the same as mimicry. Many people with Asperger’s disorder have interests like Manfred Mann’s music, or the top ten in the 1960s, which are the interests that their parents had when they were the same age as the child is now. These interests, now that we are in the twenty-first century, are eccentric. They do not develop out of the identification of the person with Asperger’s disorder with a peer group, or with parents, whose own interests have often moved on quite considerably. They are a kind of unthinking mimicry, or memorialisation, of their parents.

**Symbolic and language disorders**

It has already been noted that the current definitions of Asperger’s disorder exclude the presence of language delay. This, rather artificially, rules out adults who have residual language problems but otherwise meet all the criteria for Asperger’s disorder.

People with Asperger’s disorder may use language more than normal controls to solve cognitive problems such as facial recognition tasks. However, the words may be used in an idiosyncratic way, which in more searching tasks may prove misleading. For example, a young woman who met all Asperger’s criteria except that she had mild intellectual disability had two concepts for facial expression: narrowed eyes and round eyes. Narrowed eyes (anger, fear, disgust) worried her, but round eyes were fine. The person whose eyes were closed by plump cheeks when he or she was smiling would, of course, have been misconstrued in this scheme.

Many people with Asperger’s disorder seem to have specific arithmetic impairment (dyscalculia). Mathematics meets many of the criteria of a language and how this disorder of one type of symbolic processing is linked to language processing is unclear.