

Ants, bees, non-human primates and many other animals live in societies. Clearly, from an evolutionary viewpoint, social competence must have a neurobiological basis, of course altered by social interaction. We are all neurobiologically scripted for intuitive social interaction and are inherently social creatures. But, as expected, social competence, like any other neurocognitive function, must be a variable falling along a continuum. There are bound to be outliers. In this review we will describe three types of outlier: Asperger's syndrome will be contrasted with two similar disorders, high functioning autism and non-verbal learning disability.

HISTORY

Leo Kanner (1943) and Hans Asperger (1944), amidst a world war and a continent apart, independently described children who can best be described generically as having a 'disorder of intuitive social competence'. The autistic children described by Kanner were generally

lower functioning. Asperger's (1944) paper was written in German and, until cited by Lorna Wing in (1981), was not widely known. He described his children as 'little professors'. They were highly verbal and pedantic. Socially, they lacked intuitive ability to 'read' other individuals and to appreciate when their own compelling interests were of no further interest to their listeners.

THE RELATIONSHIP BETWEEN THE THREE DISORDERS OF INTUITIVE SOCIAL COMPETENCE

The inclusion of Asperger's syndrome in ICD-10 and DSM-IV suggests that it must have sufficiently clear diagnostic boundaries to be discriminated from high functioning autism and the more recently described disorder, non-verbal learning disability (Rourke 1989). Volkmar *et al.* 2000) importantly noted, 'with the inclusion of Asperger's disorder in both DSM IV and ICD-10, clinical use of the term

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Asperger's and related disorders

has increased dramatically. Asperger's disorder is defined, as is autism, in terms of social deficits, but early language and cognitive skills are preserved in Asperger's disorder'. Clearly there is overlap between the three disorders, but we believe there are sufficient phenomenological differences to discriminate between them.

Volkmar (2000) described this boy as a typical example of Asperger's syndrome:

Robert (not his real name), age 11 years, eight months, was seen for evaluation at the request of his parents who were concerned that despite his apparent academic skills, Robert was increasingly isolated in school. He was the younger of two children born to his parents, both physicians. Robert was born after an uncomplicated pregnancy, labour and delivery. He used his first words at one and spoke in sentences by 16 months. Motor skills were awkward and clumsy. Early on, he was an avid reader who had read the *Chronicles of Narnia* in

kindergarten. Social problems confronted Robert when he entered preschool at three years of age. He was socially inept and was seen quickly as an eccentric child. He was knowledgeable about astronomy to the point that this interest intruded on all aspects of his life. In conversation, he would shift the topic invariably or inevitably to this focus of his interest. Because of motor problems, he was evaluated by an occupational therapist. It was noted that he had 'low motor tone'. A psychiatrist suggested at eight years of age that he had an anxiety disorder. Psychological testing at 10 years of age indicated a Verbal IQ of 145, a Performance IQ of 119 and a Full Scale IQ of 135 on the WISC-III. Because of continued visual-motor coordination problems including writing, he received occupational therapy and enrolled eventually in a program for gifted children. In a psychiatric mental status examination,

Robert responded to adult greetings with appropriate, although very short, phrases. He did not respond to other people's facial expressions or gestures. He did not attend to social stimuli. He avoided eye contact and seemed to look through people. His emotional expressions lacked variability and modulation. When he described his interests in the universe, he became much more animated, but was difficult to redirect. His thinking is not disorganized. He did not appear to be depressed. He did not exhibit unusual motor behaviours. Robert, in an autobiographical statement said: 'My name is Robert Edwards. I am an intelligent, unsociable, but adaptable person. I would like to dispel any untrue rumours about me. I am not edible. I cannot fly. I cannot use telekinesis. My brain is not large enough to destroy the entire world when unfolded. I did not teach my longhaired guinea pig, Chronos, to eat everything in sight (that is the nature of the long-haired guinea)'.

An MRI scan (a T2-weighted) image, showed a 'focal hyperintensity about 1 cm in diameter without mass effect in the right middle temporal gyrus white matter just lateral to the optic radiations.

For the purposes of this review we have chosen to regard disorders of intuitive social competence as synonymous with pervasive developmental disorder (DSM-IV) and autism spectrum disorder (Rapin 1997). These are all defined by a fundamental lack of intuitive social skills believed to be secondary to weaknesses and differences in being able to perceive another individual's state of mind.

In clinical medicine it is highly desirable, but not always necessary (e.g. migraine) to define disease, disorder or syndrome by some biological marker. Without a biological marker, phenomenologically described conditions may not achieve scientific validity as distinct entities. However, behavioural scientists believe these diseases, disorders or syndromes of intuitive social competence *can* be grouped phenomenologically, and that they have both face as well as consensual validity.

The following example may help make the point. A woman says she has been sexually active, is gaining weight and is no longer having menstrual periods. In this case a valid medical diagnosis can be made using only phenomenological observations. There is predictive valid-

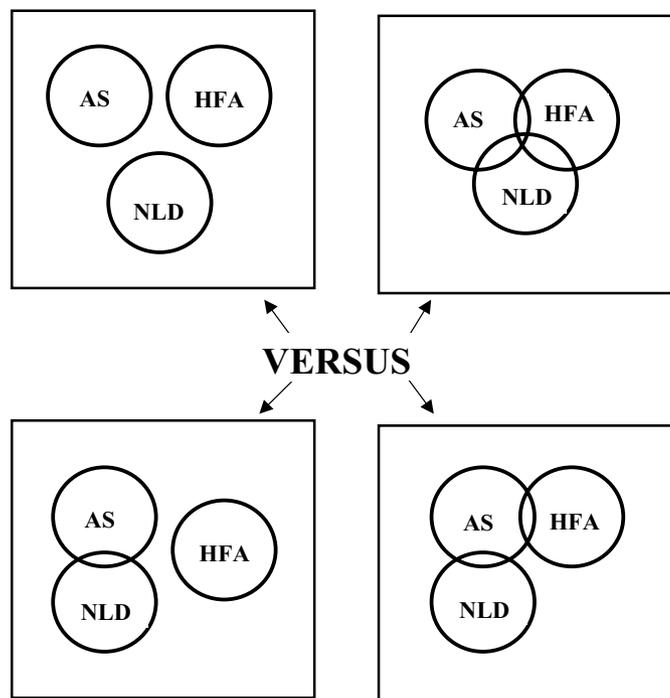


Figure 1 Various phenomenologically based views of the relationship between Asperger's syndrome, high functioning autism and non-verbal learning disability, showing that the core disorder is preserved in each. AS, Asperger's syndrome; NLD, non-verbal learning disability; HFA, high functioning autism.

ity, as well as a biological marker to externally validate the phenomenologically deduced diagnosis. Unfortunately, there are no such sharp diagnostic boundaries in disorders of intuitive social competence, but the existence of a core is virtually undisputed by most behavioural scientists. Of course, overlap at the peripheries allows considerable confusion and controversies to arise as behavioural scientists attempt to clarify the periphery, and begin to delineate subtypes. For the purposes of our discussion these contrasting subtypes are high functioning autism and non-verbal learning disability. The different ways in which they may or may not overlap is presented graphically (Figure 1), but the core disorder is preserved in each figure.

Klin *et al.* (1995) has suggested that non-verbal learning disability and Asperger's syndrome have many features in common which help discriminate them from high functioning autism. At least average verbal intellectual potential (Verbal IQ) and a significant discrepancy between intellectual potential (Full Scale IQ) and attainment on measures of non-verbal performance (Performance IQ) characterize non-verbal learning disability (Table 1). But, it is not a unitary disorder and patients have a

Table 1 Non-verbal learning disability**Discriminating features**

- At least average verbal intellectual potential
- Significant discrepancy between intellectual potential and lower performance on measures of non-verbal functioning

Consistent features

- Not a unitary disorder, heterogeneous population demonstrating a variety of levels of functioning on non-verbal tasks
- Occurs concomitantly with relative proficiency in rote verbal capacities necessary for aspects of reading and spelling performance

Variable features

- Occurs concomitantly with other handicapping conditions or extrinsic, environmental influences, but not primarily attributable to either
- Deficits in tactile perception, psychomotor coordination, and visuospatial and organizational functioning
- Deficits in aspects of complex language functioning (that is, understanding inference and humour, figurative language, language pragmatics)
- Deficits in social perception, judgement, and social interaction
- Psychiatric complications including depression

variety of levels of ability on non-verbal tasks. In contrast, relative proficiency is retained in rote verbal capacity, which is necessary for aspects of reading and spelling performance. Frequently, though not always, there are deficits in tactile perception, coordination, visuospatial and organizational functioning. The overlap with Asperger's syndrome and high functioning autism occurs because of other common though individually variable features, including deficits in complex language functions, i.e. understanding inference, humour, figurative language, and language pragmatics. Deficits in social perceptual judgements and reciprocity occur in all three disorders.

Children with high functioning autism (Table 2) must have the DSM-IV criteria, primarily impairments of social interaction such as:

- the use of non-verbal behaviours;
 - failure to develop peer relationships;
 - lack of sharing enjoyment and interest achievement;
 - lack of social emotional reciprocity.
- And impairments in the communication domain:
- delay in language development;
 - inability to initiate or sustain conversation;
 - echolalia/jargon;
 - impaired imaginative play.

Table 2 High functioning autism**Discriminating features**

- Qualitative impairment in intuitive social interactions
- Average or below average overall intelligence (Performance IQ \geq Verbal IQ)

Consistent features

- Pragmatic language disorder
- Odd stereotyped motor movements
- General delay in language development
- More often diagnosed in boys than girls
- Hyptonia (gross motor skill deficit)

Variable features

- Echolalia
- Impaired use of non-verbal behaviours
- Above average material specific visuospatial and visuomotor skills
- Hyperlexia (the precocious self-taught ability to read words with an apparent lack of comprehension)

These children also have restricted activities and interests suggested by:

- excessive preoccupations with certain interests;
- inflexible routines or rituals;
- stereotyped motor mannerisms;
- preoccupations with parts of objects.

In high functioning autism, the Full Scale IQ may be in the average range, below average or even in the retarded range (< 70). However, these measurably retarded high functioning autistic children have functional skills that clearly distinguish them from their more globally (both measurably and functionally) retarded counterparts. They are verbal – even verbose – but always in our opinion exhibit some continuing and omnipresent language disability. This disability may only affect language pragmatics (see below) and/or prosody.

Children with Asperger's syndrome (Table 3) have significantly higher overall Full Scale IQ and Verbal IQ compared with their inferior Performance IQ. Asperger's syndrome children frequently have non-verbal writing disabilities. The DSM-IV suggests that Asperger's children have qualitative impairment in social interac-

tion manifest by at least two of the four following

- impairment in the use of non-verbal behaviours to regulate social interactions;
- failure to develop peer relationships;
- lack of spontaneous seeking to share enjoyments and interests;
- lack of social or emotional reciprocity.

They also have restricted, repetitive and stereotyped patterns of behaviour, interests and activities, as manifest by at least one of the following:

- encompassing preoccupation with one or more stereotyped and restricted patterns of interest that is abnormal either in intensity or focus;
- apparently inflexible adherence to specific, non-functional routines or rituals;
- stereotyped and repetitive motor mannerisms (i.e. hand flapping, finger tapping or twisting, complex whole-body movements);
- persistent preoccupations with parts of objects.

The disturbance causes clinically significant impairment in social, occupational, or other important areas of functioning.

There is no clinically significant general delay in language (e.g. single words used by two years of age, communicative phrases used by three years of age).

There is no clinically significant delay in cognitive development or in the development of age-appropriate self-help skills, adaptive behaviour (other than in social interaction) and curiosity about the environment in childhood.

Language pragmatics

Language pragmatics mean the understanding of unspoken conversational rules and the ability to discern or express communicative intent (meaning). In Asperger's syndrome, as in high functioning autism, the use of language is impaired, notably in narrative and conversational discourse, and at a minimum pragmatics and/or prosody is affected. In our own experience, the degree of functionality or disability is related to the verbal ability (Verbal IQ).

By three years of age, children should be able to:

- request actions;
- request objects;
- make assertions;
- make denials;
- request information;

Table 3 Asperger's syndrome

Discriminating features

- Qualitative impairment of intuitive social interaction
- Above average overall intelligence (Full Scale IQ) with Verbal IQ significantly greater than Performance IQ

Consistent features

- Gross and material specific motor skill deficit
- Pragmatic language disorder (at the discourse level)
- Loquaciousness
- Restricted interests
- Age-appropriate self-help skills, adaptive
- More common in boys than girls
- No general delay of language development
- No schizophrenia
- No odd stereotyped motor movements

Variable features

- Impaired use of non-verbal behaviours
- Failure to develop peer relationships
- Lack of social or emotional reciprocity
- Inflexible adherence to rituals or routines
- Above average, material specific visuospatial and visuomotor skills (e.g. puzzles, lego)
- Hyperlexia (discrepancy between reading comprehension and word reading abilities; frequent literal interpretation)
- Features of Non-verbal Learning Disorder
- No verbal memory deficits

- request assistance;
- state information;
- appropriately use or respond to who, what, where and how questions.

But children with pragmatic language disorders often have difficulty:

- letting *others* know when they have not been understood (requesting clarification);
- responding specifically or with relevancy (tend to produce tangential comments);
- speaking clearly (or conveying information that is clear and understandable, without presupposing information on the part of the listener);
- adjusting their speech for a given context – sensitivity to listener needs;
- expressing social intentions (e.g. greetings);
- producing coherent discourse (e.g. well organized presentations of information and appropriate use of non-verbal signals);
- learning and applying the rules of socio-emotional expression;
- initiating and sustaining a conversation apart from in their own intense areas of interest.

Few formal or standardized instruments have been developed to test language pragmatics and those that are available often fail to observe individuals in a dynamic social context. Consequently, assessment is generally informal, unstructured, less than comprehensive and therefore inexact. Nonetheless, it is our experience that language pragmatics are invariably affected in Asperger's syndrome and high functioning autism. Topic maintenance is the most apparent discourse deficit in Asperger's syndrome while difficulties with social and presuppositional judgement, sensitivity to listener needs, and appreciation of context are most apparent in high functioning autism (Twachtman-Cullen 2000).

Social pragmatics

Social pragmatics broadly define the mechanisms by which human interaction occurs. A significant part of this interaction requires intact linguistic and pragmatic language skills. Social pragmatics has the same conventions (rules) as language pragmatics, but encompasses non-verbal communications. Children with disorders of intuitive social interaction competence have deficits in social pragmatics, verbal and non-verbal, which clearly distinguishes them from their intuitively socially competent peers.

Idiosyncratic behaviours

Behaviour in Asperger's syndrome and high

Language pragmatics mean the understanding of unspoken conversational rules and the ability to discern or express communicative intent (meaning)

functioning autism is frequently odd or idiosyncratic, because these individuals do not intuitively understand another person's state of mind or sense the negative reaction of others to their oddness and inappropriate behaviours. Adventitious motor movements such as tics or motor stereotypies, including persistent toe walking, hand flapping and rocking are frequently seen in high functioning autism, but, in our opinion, almost never in Asperger's syndrome or non-verbal learning disability.

Normal but odd

At the far end of the disorders of intuitive social competence continuum is what may be termed 'normal but odd'. We must all have either met or know of people who, although highly intelligent, lack social graces and may even have behavioural idiosyncrasies. Recently, a mother of a child with Asperger's syndrome who professionally teaches the gifted, reflected on the nature of her child's disability. She suggested that in her class for gifted children, even while some of them exhibited behavioural idiosyncrasies and were the subject of ridicule, they each could develop dialogue and a social bond around a common area of interest. Her own child found it difficult, if not impossible, to find individuals with whom he could share his interests for long, or at all.

NEUROPSYCHOLOGICAL AND PSYCHOLOGICAL ASPECTS

Children with Asperger's syndrome have average, above average or even superior Full Scale

IQ with Verbal IQ significantly higher than Performance IQ. In contrast, the Full Scale IQ of children with high functioning autism may be above average, average or even below. Although high functioning autism children can be superficially verbose or functionally superior, their measured IQ often falls in the retarded range. Their non-verbal skills are equal or higher than their verbal skills, irrespective of their Full Scale IQ, while in non-verbal learning disability, the Full Scale IQ is above average, or average with a big difference (often 12 or more points) between a higher Verbal IQ and a lower Performance IQ. Our clinical experience suggests, however, that non-verbal learning disability children have a range (average to above average) in Full Scale IQ in contrast to Asperger's syndrome where the IQ is generally above average, and their clinical symptomatology more closely resembles Asperger's original descriptions. There are additional treatment implications because the non-verbal learning disability children are more likely to have significant academic difficulty compared with Asperger's children.

The concept of non-verbal learning disability viewed from the standpoint of a comprehensive neuropsychological profile includes not only the significant discrepancy between Verbal and Performance IQ, but also deficits in visuospatial organization, visuoperceptual, psychomotor and non-verbal problem solving and concept-formation skills. These children have neuropsychological assets in psycholinguistic skills such as verbal learning, regular phoneme-grapheme matching, amount of verbal output and verbal classification. Major learning disabilities are seen in mechanical arithmetic and advanced levels of word recognition and spelling. There is strong evidence to suggest that virtually all the characteristics of non-verbal learning disability are seen in some children with Asperger's syndrome. These convergent characteristics, along with the more typical Asperger's symptomatology, may help distinguish these children from high functioning autism.

To repeat, Asperger's syndrome, high functioning autism and non-verbal learning disability may overlap, converge or in other ways blend but they appear to have a sound phenomenologically based core. Asperger's and high functioning autism are the most distinct and clearly separate from each other. Whether non-verbal learning disability is a 'stand alone'

entity or totally converges with Asperger's syndrome remains to be seen. We believe that the above average IQ, particularly the Verbal IQ, seen in Asperger's syndrome, contributes to the 'Little Professor' stereotype, discriminating it from non-verbal learning disability where the children are not nearly as verbose, and do not engage in excessive questioning, non-verbal learning disability children are less verbally focused and organized, and their maladaptive socio-emotional disorders are different.

GENETICS

There is general consensus that disorders of intuitive social competence have a significant genetic component, with a recurrence risk of 3–8% in families with one affected child who is autistic (Rapin 1997). In Asperger's syndrome, up to 33% of first degree relatives have 'social difficulties' (Volkmar *et al.* 1998). However, no consistent genetic or chromosomal abnormalities have been identified. There are many multiplex families, often with highly variable symptomatology. These children may even be identical or fraternal twins. We have in our practice a number of families with three (and in one family, four) affected children. Ultimately there may be no single genotype, a point suggested by the fact that there are some Down syndrome children who meet criteria for high functioning autism.

NEUROIMAGING

The findings in the case of Robert are unusual but the location is of interest, as the right middle temporal gyrus and the adjacent superior temporal sulcus play a key role in facial expression and the direction of eye gaze. Children with autism and Asperger's syndrome have head circumferences above average and at times frank macrocrania or megacephaly. The head may not be large at birth, but grows at an increasingly rapid rate. Brain volume on neuroimaging is larger overall when compared with normal children (Filipek 2000). However, routine neuroimaging is controversial – consistent abnormalities have not been found (Filipek 2000).

ELECTROENCEPHALOGRAPHY

The prevalence of epilepsy in autistic children is 7–14%. There is also a higher frequency of epileptiform abnormalities on the EEG in autistic children showing early language regression (Filipek 2000).

Children with Asperger's simply lack the capacity to articulate fears, concerns or ideas beyond a short response unless the query taps even tangentially into their area(s) of expertise

DIAGNOSIS OF ASPERGER'S SYNDROME AND THE OTHER DISORDERS OF INTUITIVE SOCIAL COMPETENCE

Asperger's children generally come to medical attention at an older age than children with other disorders of intuitive social competence. They are verbal and often academically successful. But their social ineptness, motor skill deficits and idiosyncratic interests marginalize them and they become the perfect victims. Screening questions are helpful: 'How is your child getting on socially?', 'Is your child frequently the brunt of teasing?', 'How is your child with taking turns and sharing?', 'Is your child excluded because s/he is bossy?', 'Is your child ever invited to sleepovers? The formal clinical evaluation includes intelligence testing, skilled language testing (especially pragmatics and prosody) and a measure of adaptive behaviour (e.g. Vineland Adaptive Behaviour Scales). The Australian Scale for Asperger's Syndrome or the Asperger Syndrome Diagnostic Scale can be useful. Early identification and intervention are key in this as in any other developmental disorder.

Without a biological marker, the diagnosis of Asperger's is inevitably an expert-based opinion, which relies on personal observation, parental or patient report, and patient or observer interviews.

The diagnosis of non-verbal learning disability is based primarily on a battery of neuropsychological tests (WISC-III, WRAT, and Tactile/Perceptual, Attention, Problem Solving, Motor/Psychomotor assessments). Neurolinguistic testing is an essential complement. A psychiatric/psychological evaluation is also

important because of the frequent concurrent psychiatric disorders, including severe depression.

The diagnosis of high functioning autism may be made by the use of parent-teacher report or observation rating scales. In-depth discussions of these can be found in Filipek (2000).

Recommended instruments include:

- parent interviews;
- The Gilliam Autism Rating Scale;
- The Parent Interview for Autism;
- The Pervasive Developmental Disorders Screening Test Stage 3;
- The Autism Diagnostic Interview.

The diagnostic observation instruments include:

- The Childhood Autism Rating Scale;
- The Screening Tool for Autism in Two Year Olds;
- The Autism Diagnostic Observation Schedule – Generic.

These instruments should enable clinicians to determine whether diagnostic criteria have been met. This diagnostic standard is therefore criterion-based. Tables 1–3 propose a discriminator based system. Identifying biological markers in these disorders would be the preferred approach to diagnosis. However, from a teaching/understanding point of view, even if phenomenologically based, these tools can still be useful. They should be viewed as a best effort given the 'state-of-the-art' and are therefore only a point of departure. There are bound to be revisions as more knowledge emerges.

MANAGEMENT

General

Because children with Asperger's are so verbal, teachers, parents and others assume they can be reasoned with using verbally mediated questions and answers. They cannot understand why the most common response to any 'who', 'why', 'where', 'when' or 'how' questions is, 'I don't know.' When one recalls that these children lack intuitive pragmatic language skills, their response is understandable. Children with Asperger's simply lack the capacity to articulate fears, concerns or ideas beyond a short response unless the query taps even tangentially into their area(s) of expertise. Because they do not know how to respond they may be accused of not paying attention, being unconcerned, oppositional or defiant. The oppositional-defiant response behaviour soon generalizes to all behaviours to

the frustration of caregivers and teachers. Medications, often multiple and in increasingly large doses, are expected – wrongly – to impact on what is essentially a communication problem.

Therefore, social learning must use an operant behaviour consequence approach. Lacking intuitive social skill, the Asperger's child must be taught using a rule-based approach be it for purposes of discourse or social intervention, such as the concept of personal space or appropriate touching. Civilized society has numerous rules, many of which are enforced without explanation. A rule-based method in a natural setting simply emphasizes this approach over one relying on insight. Human nature usually compels caregivers to provide a rationale for their requests or demands. While it may be crass to say so, unfortunately this is often literally a waste of time and breath with Asperger's children. Compassionate firm and consistent rule setting is difficult but necessary. 'You broke the rule and you know the consequences that you can expect.' Many children with Asperger's syndrome enjoy rules just as much as they enjoy rituals and preservation of sameness. Invariably, these rules will take on a life of their own, and the child's focus will shift to the rule rather than the rule maker. This allows the rule maker to escape being seen as the villain.

Language (verses speech) therapy should focus on developing pragmatic language skills, if need be employing a rote methodology. Psychological counselling should emphasize setting standards for social interaction rather than relying on an insight-orientated approach. Family involvement is critical.

Education

At school, we have found that providing an Asperger's child with an individual adult trainer, mentor or shadow is important initially. This individual should have special training in working with these children. The services of this individual can be faded out as the child adjustment improves. In later school years, ridicule increases and becomes, in our experience, almost totally impossible to control. This, with 'change' of any kind, leads to great anxiety. The ultimate solution may be a small school with an excellent staffing/pupil ratio. This setting is essentially a 'club' where acceptance is guaranteed and often includes other children with the same disability. The last place these emotionally vulnerable children should be is in a class for emotionally or

behaviourally disturbed children. An Asperger's child becomes in this context, the perfect victim with skilled victimizers.

Pharmacotherapeutic agents

There is no traditional or non-traditional remedy or cure for Asperger's syndrome. Pharmacotherapeutics do not target the primary deficit but can be used as adjunctive agents. The stimulants (e.g. methylphenidate, dextroamphetamine) can enable the child to better attend or focus and thus indirectly improve behaviour. An agent such as clonidine can be used for anxiety, pre-emptively or for sleep. In this situation it appears to have a predictable response.

Neuroleptics can be used in extreme situations, but the long-term risk of tardive dyskinesia limits their usefulness. Selective serotonin reuptake inhibitors and antiepileptic agents can be used as 'mood stabilizers'. In our experience these agents have an inconsistent effect and a dramatic response is very infrequent. It is particularly important in these children to 'start (very) low and go (very) slow.' They do not adjust well to any changes and this may account in part for exaggerated responses to even the smallest dose of any medication.

Parent and patient information

[Http://yale.med.info/chidstdy/autism](http://yale.med.info/chidstdy/autism)

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