

Editorial

## What's so special about Asperger Syndrome?

### 1. A brief history

In the field of autism, Asperger Syndrome (AS) is a relatively late arrival. It has become almost ironic that the two subtypes of classic autism and AS were described almost simultaneously by two independent clinicians (Asperger, 1944; Kanner, 1943).

Kanner was born on June 13th 1894 in Klekotow, Austria. He left his home country for the US in 1924, moving to John Hopkins University School of Medicine in 1930 to set up the country's first child psychiatry service within a paediatric hospital. His rich descriptive prose was codified (Rutter, 1978), becoming the basis of subsequent definitions of autism around the world. Kanner died on April 4th 1981. The subgroup that he described is variously known as 'classic autism,' 'Kanner-type autism,' or 'autistic disorder.' It bears the hallmarks of social difficulties, repetitive behaviour, unusually narrow interests, language, and communication delays, with a large risk of (what in the UK is called) learning disability or (what in the US is still called) 'mental retardation'—that is, general developmental delay with below average IQ.

Hans Asperger was director of the University Children's Clinic in Vienna. He was born on February 18th 1906 in Vienna, joined the Youth Movement in the 1920s and became a medical doctor in 1931, staying in Austria throughout his career. He therefore wrote his account in German. Although Asperger's work was described in the first issue of the *Journal of Autism and Child Schizophrenia* (van Krevelen, 1971), it remained virtually unknown outside German-speaking countries. He died on October 21, 1980.

Lorna Wing is recognized as having drawn the English-speaking medical community to the existence of AS by summarizing Asperger's observations and providing clinical illustrations in what became an influential journal article (Wing, 1981). The first systematic studies began to appear in the late 1980's in the UK (Tantam, 1988), Sweden (Gillberg & Gillbert, 1989), and North America

(Szatmari, Bartolucci, & Bremner, 1989). The use of the term Asperger Syndrome in both clinical practice and research grew exponentially with its formalization in DSM-IV and ICD-10 (Volkmar et al., 1994). Nevertheless, questions about its validity (e.g., relative to autism in individuals with normative IQ) persist to this day, a vexing problem compounded by the use of different definitions in different studies (Klin, Pauls, Schultz, & Volkmar, 2005). The subgroup called Asperger Syndrome is typically seen today as lying on the same autistic spectrum as classic autism because these individuals also have the social difficulties, repetitive behaviour, and unusually narrow interests. Like autism, it is considered to be a developmental condition of early onset. But unlike Kanner-type autism, there is no history of language delay or general developmental delay.

It was not until Uta Frith's translation of Hans Asperger's original paper (Frith, 1991) that his work became widely available to English-speaking clinical researchers, parents and advocates around the world. The important point in this brief history is to emphasize that it is less than 15 years since the first book in English on the topic of AS, and less than 11 years since its inclusion in formal diagnostic manuals that confer special education entitlements and inform the medical and mental health community at large. Prior to that time, AS was virtually unknown. Today it is recognized as a common subgroup on the autistic spectrum, and there are hundreds of books, articles, and websites devoted to describing it, documenting the unique needs and assets of this population and their families. During the same period of the 1990's and early 21st century, prevalence figures of the autistic spectrum diagnoses have increased dramatically from 4 per 10,000 to maybe 4–5 per 1000 (Baird et al., 2000; Chakrabarti & Fombonne, 2001), representing a momentous increase in the number of individuals being identified to have autism or a variant of it. AS is recognized as an important subgroup in this spectrum (Fombonne & Tidmarsh, 2003).

## 2. Why a special issue on Asperger Syndrome?

As editors, we saw value in a special issue of *Brain and Cognition* for several reasons.

First, although there has been an explosion of writing about AS, much of this has been clinical or anecdotal or family or first person perspectives, but there have been relatively few well-controlled experimental studies looking at either aspects of brain or cognition in people with AS, distinct from classic autism.

Second, AS, more than perhaps almost any other developmental disability, highlights the need for research to consider not only the deficits displayed by individuals with this condition but also, and importantly, their strengths and even special talents (Baron-Cohen, 2000). It is consensually agreed that appropriate programs of intervention address the individuals' needs while capitalizing on the individual's assets (National-Research-Council, 2001). The latter are as important as the former, since more positive prospects for an independent and fulfilling adult life will often depend on the extent to which talents are nurtured into vocational avenues. The studies reported here throw light on the nature of the deficits in the condition, but alongside these there are areas of not just intact performance, but also of unique skills.

Third, AS is a complex condition in the familiar sense that to understand it requires an analysis at many different levels, from behaviour to cognition to neurobiology to genetics. Thus, for example, autism involves atypical brain function and structure in neural substrates that subserve socialization, including the amygdala (Baron-Cohen et al., 2000) and closely and densely interconnected mid-temporal and medial prefrontal cortex (Schultz et al., 2000). It involves rapid early brain growth (Courchesne, Carper, & Akshoomoff, 2003), and mechanisms that may lead to abnormal neural architecture have been proposed (Casanova, Buxhoeveden, Switala, & Roy, 2002). It will be important to test if these characteristics apply to the whole range of manifestations within the autistic spectrum including AS. Equally, a strong genetic component in classic autism has been demonstrated by a number of studies showing an increased concordance rate in monozygotic (identical) (MZ) compared to dizygotic (non-identical) (DZ) twins as well as an increased risk to siblings, relative to the general population prevalence (Folstein & Rutter, 1977; Steffenburg et al., 1989). Bolton et al. (1994) and others also demonstrated high rates of autism in the siblings of autistic probands as well as increased rates of cognitive and social abnormalities in their first-degree relatives, compared to relatives of controls. Advances in the understanding of the molecular basis of this genetic liability have increased in pace and scope in the past 3 or 4 years (Veenstra-Vanderweele & Cook, 2004). Although classic autism and AS may occur within the same sibship (Burgoine & Wing, 1983) and most researchers would agree on shared genetic mechanisms (Volkmar, Klin, & Pauls, 1998), there has not, as yet, been a twin study of AS. This lag in knowledge is no longer acceptable, particularly considering that

autism spectrum conditions are the most strongly genetic conditions among the neurodevelopmental conditions (Bailey et al., 1995).

Estimates of broad heritability for classical autism are of the order of 80–90%. A number of lines of evidence point to autism being a polygenic complex condition and Risch et al. (1999) proposed that the underlying inheritance is most likely to be reflected in an epistatic model and suggested that up to 15 genes may be involved. Although AS is regarded as an autistic spectrum condition, its genetic underpinnings are less well established largely because most studies have focused on classic autism. Gillberg (1989) has demonstrated a number of pedigrees showing familiarity of Asperger Syndrome. The most strongly implicated chromosomal regions harbouring susceptibility genes for autism are 7q (Barrett et al., 1999; IMGSAC, 1998, 2001), 2q (Buxbaum et al., 2001; IMGSAC, 2001) as well as 15q (Cook et al., 1997; Phillippe et al., 1999), although this literature is updated almost monthly. In addition there is an increasingly large literature focussing on candidate gene analysis in autism. A number of genetic variants have been studied which have demonstrated association with autism. However, the majority of findings have not been widely replicated. Molecular genetic studies specifically examining AS are only beginning to emerge. A recent linkage genome scan of AS (Yisaukko-oja et al., 2004) reported several genomic regions of interest. It will be important to study if the genetics and neurobiology of AS overlaps with that found in classic autism. If so, our attention may then move to possible mediators of outcome that could elucidate the wide manifestations of autistic spectrum conditions, and pertinent mechanisms leading to phenotype variability.

In this special issue, some of the studies reveal that AS involves different sensitivity thresholds in sensory processing (Blakemore et al., 2006), difficulties in multi-tasking (MacKinlay, Charman, & Karmiloff-Smith, 2006), executive skills (Happe, Booth, Charlton, & Hughes, 2006), and emotional facial expression recognition (Ashwin, Wheelwright, & Baron-Cohen, 2006), as well as contrasting performance in social versus physical attribution skills (Klin & Jones, 2006). These difficulties can be contrasted to intact skills in some areas of performance (White, Hill, Winston, & Frith, 2006). Other studies show atypical neural activation patterns using fMRI (Harris et al., 2006) or ERP (Henderson et al., 2006). Finally, two genetic-MRI designs reveal differences in brain structure even within monozygotic twins with AS (Belmonte & Carper, 2006) and in parents of children with this condition (Baron-Cohen et al., 2006).

The aim of this Special Issue is to encourage more research into AS, so that we can overcome nosologic debates (Volkmar & Klin, 2005) and eventually have answers to such fundamental questions as “How does AS differ from autism?” and “How does AS differ from typical development?” In tandem, the aim is that such research will generate new ways of thinking about intervention, so that

people with AS can find the right support for their difficulties whilst being respected for their differences and valued for their special qualities.

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