Asperger's Syndrome

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DESCRIPTION

Asperger's syndrome represents an autistic spectrum disorder that is primarily characterized by deficits in social reciprocity, diminished nonverbal communication skills, restricted or circumscribed interests, and repetitive and/or stereotyped behaviors. In comparison, autism is also associated with verbal language delays, whereas Asperger's syndrome is not.

Asperger's syndrome was first described by Hans Asperger, a Viennese pediatrician, in 1944, who published a clinical description of a group of children who shared an unusual combination of developmental weakness and strengths. These children had severe social and communication abnormalities, restricted interests, motor delays, and clumsiness. Asperger initially named this syndrome autistic psychopathy (Miller & Ozonoff, 2000). However, although he noted the aforementioned deficits, this group of children acquired language at normal or earlier age and had normal intelligence and some specific strong abilities such as visuospatial skills and verbal abstraction abilities. In 1943, the child psychiatrist Leo Kanner published a description of a group of children with similar characteristics but with no mention of motor deficits as a core symptom of the syndrome.

NEUROPATHOLOGY/PATHOPHYSIOLOGY

The neuropathological basis of Asperger's syndrome remains unclear. Studies have assumed that Asperger's syndrome is part of the autism spectrum disorders, and the causes and mechanisms to explain the symptoms would be essentially the same and the differences would be attributable to the severity (Wicker et al., 2008). Overall, it is accepted that Asperger's syndrome is a polygenetic disorder, and several neurological findings of children with autism spectrum disorder have been described including deficits in intrahemispheric neural connectivity, reductions in the size of the corpus callosum, reductions in the inhibitory cortical activity, abnormal development of gray and white matter, increased activation in Wernicke's area, diminished activation in Broca's area during cognitive processing, decreased connectivity between the inferior frontal gyrus and the limbic system, and a significant deficit in the connectivity between the primary sensory cortex and the association cortex (Minshew & Williams, 2007). In summary, more recent findings support the hypothesis of Asperger's syndrome as a large-scale neural systems disorder in which deficits in cortical connectivity and abnormal balance between excitatory and inhibitory mechanisms might play the main role. For instance, one of the mechanisms that explain the social interaction impairment found in autism spectrum disorders is the deficit in emotional processing of face expressions. This weakness can be explained by the lack of neural connectivity (Wicker et al., 2008). Wicker et al. (2008) in a recent publication found that adults with autism spectrum

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Asperger's syndrome
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disorder, when observing angry and happy faces, have an abnormally low activation of the areas involved in the processing of high-order emotional information: dorsomedial prefrontal cortex (DMPFC) and right ventrolateral prefrontal cortex. However, normal activity was found in the areas that are usually activated during perceptual analysis of facial features and expressions such as superior temporal sulcus and fusiform gyrus. This lack of synchrony in the activation of these areas and the deficit of signals from the amygdala to the DMPFC might explain the failure in relating the emotional features of faces with the initiation of socially appropriate behaviors.

NEUROPSYCHOLOGICAL/CLINICAL PRESENTATION

Early language development delays are not expected in children with Asperger's syndrome (Miller and Ozonoff, 2000). In fact, earlier development of language, even before children walk appears to be common. However, later during preschool years some abnormalities may appear such as echolalia, pronoun reversal, repetitive speech, low speech initiative, and limited play. During adulthood, language can be stilted, gauche, or pedantic. Individuals may demonstrate an odd use of words while also demonstrating idiosyncratic gestural accompaniments (Perlman, 2000). The Boston Naming Test, the Rapid Automatized Naming Test, and the Verbal Fluency Test have been used to assess language (Saalasti et al., 2008).

Motor impairment is present even in children with no mental retardation measured by the Movement Assessment Battery for Children (Green et al., 2002), and performance is also poor in tests of apraxia, one-leg balance with eyes closed, tandem gait, and repetitive finger thumb apposition. However, no differences on tests of finger tapping, grooved pegboard, trail making, or visual integration have been found (Weimer, Schatz, Lincoln, Bellantyne, & Trauner, 2001).

Overall, cognitive abilities appear to be normal with higher scores in verbal scales of the Wechsler Intelligence Scale for Children in comparison with nonverbal/performance scales (Miller & Ozonoff, 2000). Consequently, wide variability in cognitive profiles of children with Asperger's syndrome is not uncommon and, in turn, cognitive giftedness is rare (Chiang & Lin, 2007; Edgin & Pennington, 2005). Some memory problems can be observed during free recall tasks, but the deficit disappears when cues are provided (Bowler, Gardiner, & Berthollier, 2005). Creativity and imagination are impoverished when using a test of imaginative fluency (Craig & Baron-Cohen, 1999). Executive functioning is the area more affected in this syndrome as evidenced for defective performance in tasks of response selection/inhibition, flexibility, planning, working memory, abstract problem solving, visual memory, and multitasking (Christ, Holt, White, & Green, 2007; Edgin & Pennington, 2005; Happé, Booth, Charlton, & Hughes, 2006; Hill & Bird, 2006).

The social challenges that individuals with Asperger's syndrome have seem to be related with their inability to attribute mental states to themselves and others in order to explain and anticipate behaviors. This cognitive skill has been called “theory of mind” (Duverger, Da Fonseca, Bailly, & Deruelle, 2007). The social interaction impairment involves deficits in empathy, poor eye contact, isolation, physical contact defensiveness, and generally inappropriate social behavior. These features as well as other clinical characteristics are successfully assessed by different instruments such as the Autism Diagnostic Interview-Revised, the Autism Diagnostic Observation Schedule (Miller & Ozonoff, 2000), and the Autism Spectrum Screening Questionnaire (Saalasti et al., 2008).

DIAGNOSIS

Diagnosis of Asperger's syndrome is largely dependent upon clinical interview and developmental history. In addition, neuropsychological and psychological assessment, both with performance-based measures and self-report or observer rating, may offer further insight into the diagnostic picture. Today, modern systems of diagnosis such as the *Diagnostic and Statistical Manual of Mental Disorders* (*DSM-IV*, American Psychiatric Association [APA], 2000) and the International Classification of Diseases (ICD) (Isaac, Janca, & Sartorius, 1994) place Asperger'& Sartorius, 1994) place Asperger's syndrome in the category of Pervasive Developmental Disorders. Both diagnostic systems define this syndrome with similar features that were earlier noticed by Leo Kanner and Hans Asperger, including impairment in social interaction and restricted and stereotyped patterns of behavior, interests, and activities. However, the *DSM-IV* requires the criteria of absence of clinical delay in language, cognitive development, and self-help skills to make the diagnosis. The diagnosis of autistic disorder has to be assigned when these cognitive delays and deficits of adaptive skills are present.

Besides the differences in the diagnostic criteria that exist between Asperger's syndrome and autism, both the *DSM-IV* and *ICD-9* point out that there is an overlap among symptoms and features. As such, children with autism who demonstrate high performance in social and cognitive skills can be easily misdiagnosed with Asperger's syndrome. Miller and Ozonoff (2000) explored the external validity of Asperger's disorder by comparing a group of children previously diagnosed with high-functioning autism with another group of children with Asperger's syndrome. In their research, they used clinical measures of autism, intelligence, motor skills, visuoperceptual skills, executive functioning, and cognitive flexibility. The results showed that both groups were not different in most of the measures and the variable that explained the differences between groups was intellectual ability. Overall, the group of children with diagnosis of Asperger's syndrome performed better than children with autism only in intellectual ability. Coinciding with the idea that the difference between these two disorders accounts for severity of symptoms, one approach that is being adopted is to treat autism and Asperger's disorders as being part of a continuum or a spectrum of symptoms that varies in severity. Consequently, children with low language, cognitive, intellectual, and adaptive skills would be located on one side of the spectrum. In the other more adaptive extreme of the spectrum, children with Asperger's syndrome and high-functioning autism would be placed. This approach has been challenged by several publications that found important variability in cognitive and adaptive profiles of both groups (i.e., high-functioning autism and Asperger's syndrome) yet only few essential differences (Chiang & Lin, 2007). These differences between Asperger's syndrome and high-functioning autism remain controversial, and it is suggested that high-functioning autism and Asperger's syndrome would be the same disorder. Moreover, the differences that have been found, appear to be minor or only based on

clinical symptoms profiles from a clinical perspective (Paul, Orlovski, Marcinko, & Volkmar, 2008; Schultz et al., 2000). Williams, Goldstein, Kojkowski, and Minshew (2008) when studying IQ profiles of high-functioning autistic children with nonverbal learning disabilities suggested that clinical scales such as the Autism Diagnostic Observation Schedule are more reliable and accurate than IQ profiles in diagnosing autism. Similarly, Spek, Scholte, and Van Berckelaer-Onnes (2008) found that differences between verbal IQ and performance IQ cannot help differentiate adults with Asperger's syndrome from high-functioning autism even though the Asperger's syndrome group performed slightly better in verbal IQ than the autistic subjects. The same difference is found when comparing Asperger's disorder with pervasive developmental disorder not otherwise specified (Koyama & Kurita, 2008).

TREATMENTS

Most of the treatments have been focused on social skills development, even in the school setting (Williams, Keoning, & Scahill, 2007). However, psychotherapy in adults with this disorder is also possible as reviewed by Ramsay, Brodkin, Cohen, Listerud, and Rostain (2005). This review describes how social skills groups and cognitive behavior-based interventions can be useful not only in treating frequent comorbid conditions such as depression and anxiety but also negative beliefs and social aversions that are common in these individuals. Pharmacotherapy is also useful in treating comorbid obsessive-compulsive, mood, sleep, and anxiety problems. The families of the children with this disorder should be assessed for therapeutic and psychosocial interventions as evidenced by the high percentage of mothers who are at higher risk of physical and mental illnesses because of the high stress that they face in their role as caregivers (Allik, Larsson, & Smedje, 2006).

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