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Review Article

Asperger's Syndrome: A timely update

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Abstract

Asperger's syndrome is a developmental disorder that affects a person's ability to socialize and communicate effectively with others. Children with Asperger's syndrome typically exhibit social awkwardness and an all-absorbing interest in specific topics. In this review a systematic search of Asperger's syndrome including its symptoms, epidemiology, pathophysiology, diagnosis and treatment was carried out focusing on literatures published in scientific databases like PubMed, Scopus, Web of Science, Medline, Google Scholar etc. In this review, we tried to give a timely and comprehensive update about Asperger's syndrome to reveal therapeutic and gaps requiring future research opportunities.

Introduction

Asperger's syndrome is a milder variant of Autistic Disorder. Both Asperger's syndrome and Autistic Disorder are in fact subgroups of a larger diagnostic category. This larger category is called either Autistic Spectrum Disorders, mostly in European countries, or Pervasive Developmental Disorders (PDD), in the United States. In Asperger's syndrome, affected individuals are characterized by social isolation and eccentric behavior in childhood. There are impairments in two-sided social interaction and non-verbal communication. Though grammatical, their speech may sound peculiar due to abnormalities of inflection and a repetitive pattern. Clumsiness may be prominent both in their articulation and gross motor behavior. They usually have a circumscribed area of interest, which usually leaves no space for more age appropriate, common interests. Some examples are cars, trains, French Literature, doorknobs, hinges, cappuccino, meteorology, astronomy or history.

Asperger's syndrome (AS) is an autism spectrum disorder. It is milder than autism but shares some of its symptoms. It is more common in boys than girls. An obsessive interest in a single subject is a major symptom of AS. Some children with AS have become experts on dinosaurs, makes and models of cars, even objects as seemingly odd as vacuum cleaners. Their expertise, high level of vocabulary and formal speech patterns make them seem like little professors. Children with AS have trouble reading social cues and recognizing other people's feelings. They may have strange movements or mannerisms. All of these make it difficult for them to make friends. Problems with motor skills are also common in children with AS. They may be late learning to ride a bike or catch a ball, for example. Treatment focuses on the three main symptoms: poor communication skills, obsessive or repetitive routines, and physical clumsiness.

Asperger's syndrome is named after Hans Asperger, an Austrian pediatrician who, in 1943, published a paper describing Autism, and then, in 1944, a paper about what was later to be called Asperger's syndrome. At the same time, Leo Kanner, an American psychiatrist who emigrated from Austria, published a

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paper describing Early Infantile Autism, sometimes also referred to as Kanner's Syndrome.

Although there are key physiological differences between the brains of people with Autism and those of us with Asperger's syndrome, at this time diagnosis is made on the basis of observable behaviors, and in this respect the two differ by: onset is usually later, outcome is usually more positive, social and communication deficits are less severe, circumscribed interests are more prominent, verbal IQ is usually higher than performance IQ (in autism, the case is usually the reverse), clumsiness is more frequently seen, family history is more frequently positive, neurological disorders are less common.

While sharing many of the same characteristics as other Autism Spectrum Disorders (ASD's) including Pervasive Developmental Disorder - Not Otherwise Specified (PDD-NOS) and High-Functioning Autism (HFA), AS has been recognized as a distinct medical diagnosis in Europe for almost 60 years, but has only been included in the U.S. medical diagnostic manual since 1994 ("Asperger Disorder" in the DSM-IV).

Individuals with AS and related disorders exhibit serious deficiencies in social and communication skills. Their IQ's are typically in the normal to very superior range. They are usually educated in the mainstream, but most require special education services. Because of their naiveté, those with AS are often viewed by their peers as "odd" and are frequently a target for bullying and teasing.

They desire to fit in socially and have friends, but have a great deal of difficulty making effective social connections. Many of them are at risk for developing mood disorders, such as anxiety or depression, especially in adolescence. Diagnosis of autistic spectrum disorders should be made by a medical expert to rule out other possible diagnoses and to discuss interventions.

Characteristics of Asperger's Syndrome

Each person is different. An individual might have all or only some of the described behaviors to have a diagnosis of AS. These behaviors include the following:

- ✓ Marked impairment in the use of multiple nonverbal behaviors such as: eye gaze, facial expression, body posture and gestures to regulate social interaction.
- ✓ Extreme difficulty in developing age-appropriate peer relationships (e.g. AS children may be more comfortable with adults than with other children).
- ✓ Inflexible adherence to routines and perseveration.
- ✓ Fascination with maps, globes, and routes.
- ✓ Superior rote memory.

- ✓ Preoccupation with a particular subject to the exclusion of all others. Amasses many related facts.
- ✓ Sensitivity to the environment, loud noises, clothing and food textures and odors.
- ✓ Speech and language skills impaired in the area of semantics, pragmatics and prosody (volume, intonation, inflection, and rhythm).
- ✓ Difficulty understanding others' feelings.

Epidemiology of Asperger's syndrome

In a total population study of children between ages 7-16 in Goteborg, Sweden, minimum prevalence of Asperger's syndrome was 36/10,000 (55/10,000 of all boys, and 15/10,000 of all girls), and the male/female ratio was 4:1. The prevalence of autism has traditionally been estimated around 4-5/10,000. A recent study from United Kingdom found the prevalence of autism at 17/10,000, and the prevalence of all Autistic Spectrum Disorders (including autism) at 63/10,000.

Pathophysiology of Asperger's syndrome

Despite the now widely accepted fact that biological factors are of crucial importance in the etiology of autism, so far the brain imaging studies have shown no consistent pattern, no consistent evidence of any type of lesion and no single location of any lesion in subjects with autistic symptoms. This inconsistency in the results of various brain imaging studies has been attributed to the fact that people with autism represent a highly heterogeneous group in terms of underlying pathology. Therefore there is an ongoing effort to specify more homogenous subgroups among autistic individuals to enhance the accuracy of etiologic inquiry. This approach has been supported with the inclusion of the diagnosis 'Asperger's syndrome' in the Fourth Edition of the Diagnostic and Statistical Manual of Mental Disorders (DSM-IV) of the American Psychiatric Association.

Associated medical conditions such as fragile-X syndrome, tuberous sclerosis, neurofibromatosis and hypothyroidism are less common in Asperger's syndrome than in classical autism. Therefore it may be expected that there are fewer major structural brain abnormalities associated with Asperger's syndrome than with autism. To our knowledge, a very small number of structural brain abnormalities have been so far associated with Asperger's syndrome, which includes left frontal macrogyria, bilateral opercular polymicrogyria and left temporal lobe damage. On the other hand brain imaging techniques like positron emission tomography and single photon emission tomography which provides information about the functional status of brain may be more helpful in determining the brain dysfunction in

individuals with Asperger's syndrome. Detailed neuropsychological testing may support these findings providing information about the performances of individual right or left hemispheric brain regions. The first SPECT study in a patient with Asperger's syndrome was published by the host of this page and his colleagues and found left parietooccipital hypoperfusion. Continuation of research in Asperger's syndrome with various brain imaging techniques in coordination with neuropsychological evaluation in larger samples is clearly needed in this area.

Diagnosis of Asperger's syndrome

DSM-IV Diagnostic Criteria for Asperger's syndrome:

According to this, qualitative impairment in social interaction, as manifested by at least two of these: marked impairment in the use of multiple nonverbal behaviors such as eye-to-eye gaze, facial expression, body postures, and gestures to regulate social interaction, failure to develop peer relationships appropriate to developmental level, lack of spontaneous seeking to share enjoyment, interests, or achievements with other people (e.g., by a lack of showing, bringing, or pointing out objects of interest to other people), lack of social or emotional reciprocity.

Restricted repetitive and stereotyped patterns of behavior, interests and activities, as manifested by at least one of these: 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, nonfunctional routines or rituals, stereotyped and repetitive motor mannerisms (e.g., hand or finger flapping or twisting, or complex whole-body movements), persistent preoccupation 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 age 2 years, communicative phrases used by age 3 years). There is no clinically significant delay in cognitive development or in the development of age-appropriate self-help skills, adaptive behavior (other than in social interaction), and curiosity about the environment in childhood. Criteria are not met for another specific Pervasive Developmental Disorder or Schizophrenia.

Gillberg's diagnostic criteria for Asperger's syndrome:

According to it severe impairment in reciprocal social interaction includes at least two of these: inability to interact with peers, lack of desire to interact with peers, lack of appreciation of social cues, socially and emotionally inappropriate behavior. All-absorbing narrow interest includes at least one of these: exclusion of other activities, repetitive adherence, more wrote than meaning. Imposition of routines and

interests include at least one of these: on self, in aspects of life, on others. Speech and language problems include at least three of these: delayed development, superficially perfect expressive language, formal, pedantic language, odd prosody, peculiar voice characteristics, impairment of comprehension including misinterpretations of literal/implied meanings. Non-verbal communication problems include at least one of these: limited use of gestures, clumsy/gauche body language, limited facial expression, inappropriate expression, peculiar, stiff gaze.

Asperger's syndrome may not be the only psychological condition affecting a certain individual. In fact, it is frequently together with other problems such as: Attention Deficit Hyperactivity Disorder (ADHD), Oppositional Defiant Disorder (ODD), Depression (Major Depressive Disorder or Adjustment Disorder with Depressed Mood), Bipolar Disorder, Generalized Anxiety Disorder, Obsessive Compulsive Disorder (OCD).

Treatment of Asperger's syndrome

There is no specific treatment or cure for Asperger's syndrome. All the interventions outlined below are mainly symptomatic and/or rehabilitational. Psychosocial interventions include individual psychotherapy to help the individual to process the feelings aroused by being socially handicapped, parent education and training, behavioral modification, social skills training and educational interventions. Psychopharmacological interventions includes psycho stimulants (methylphenidate, dextroamphetamine, met amphetamine), clonidine, tricyclic antidepressants (desipramine, nortriptyline), Strattera (atomoxetine) for hyperactivity, inattention and impulsivity. Mood Stabilizers (valproate, carbamazepine, lithium), beta blockers (nadolol, propranolol), clonidine, naltrexone, neuroleptics (risperidone, olanzapine, quetiapine, ziprasidone, haloperidol) are given for irritability and aggression. For preoccupations, rituals and compulsions, SSRIs (fluvoxamine, fluoxetine, paroxetine) and tricyclic antidepressants (clomipramine) are used. SSRIs (sertraline, fluoxetine), tricyclic antidepressants (imipramine, clomipramine, nortriptyline) are preferred for anxiety.

Conclusion

The outcome for children with Autism Spectrum Disorders is related to intellectual functioning and communication skills. Children with normal or above normal intelligence and normal or near normal speech and language often finish high school and attend college. Although difficulties with social interaction and awareness may persist, they can often

do well in specific work settings and develop lasting relationships with family and friends. Access to ongoing counseling, support and assistance increases the likelihood of a positive and successful outcome. The timely and comprehensive update about Asperger's syndrome in this review is helpful to reveal therapeutic and gaps requiring future research opportunities.

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