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Attention deficit disorder with or without hyperactivity or impulsivity in children with Down's syndrome

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Abstract

Children with Down's syndrome show a higher prevalence of attention deficit disorder with or without hyperactivity or impulsivity (ADHD) than the rest of the general population. The diagnosis and identification of ADHD is important because it can affect performance at school and cause behavioural disturbances.

This research study has two objectives. First of all, in this review we consider the repercussions that ADHD has on Down's syndrome children. Secondly, we present a systematic analysis of the articles published in the scientific literature relating to the tests used to diagnose ADHD in DS children.

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Trastorno por déficit de atención con o sin hiperactividad en los niños con síndrome de Down

Resumen

Los niños con síndrome de Down tienen una prevalencia más elevada que el resto de población general de presentar trastorno por déficit de atención con o sin hiperactividad o impulsividad (TDAH). El diagnóstico e identificación del TDAH es importante porque puede afectar el rendimiento escolar y causar trastornos de la conducta.

El objetivo de este trabajo es doble. En primer lugar, se considera en esta revisión la repercusión del TDAH en los niños con síndrome de Down. En segundo lugar, se presenta un análisis sistemático de los artículos publicados en la bibliografía científica relativos a los tests utilizados para el diagnóstico de TDAH en niños con síndrome de Down.

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Attention deficit disorder with or without hyperactivity or impulsivity and Down's syndrome

Attention deficit disorder with or without hyperactivity or impulsivity (ADHD) affects approximately 3%-7% of schoolchildren in the general population,¹ whilst in mentally handicapped children this figure is as high as 14.8%² and, in Down's syndrome (DS), it can reach 9%.³ The cardinal symptoms are lack of attention, hyperactivity and impulsivity. The parents of children and adolescents with DS have described more behavioural disturbances and problems related with attention when they compare them with siblings who have shown normal development.⁴ Greater hyperactivity has also been described in DS children from 5-11 years of age when they are compared with a sibling group.⁵

ADHD is a genetic neurobiological disorder. In this disorder, there are functional and anatomical changes in the prefrontal cortex and its connections with the basal nuclei (especially the striate nucleus) and the cerebellum, predominantly involving catecholaminergic pathways and their neurotransmitters, dopamine and noradrenaline.

In a recent study conducted at the University of Cardiff,⁶ it was demonstrated that children with ADHD exhibit a greater number of DNA deletions or duplications, which are known as copy number variations (CNV), in comparison with children who do not have this syndrome. CNV cause the genes which are found in affected regions to be more or less active than they ought to be, leading to the production of excessive or insufficient amounts of the proteins which they code for. It has been postulated that, depending on the genes affected, children with ADHD will manifest one or other subtype of the disorder and, depending on the number of copies of each gene, ADHD intensity will vary.

There are two important concepts which can help us to understand how ADHD and DS are related: dual diagnosis and behavioural phenotype.

The term dual diagnosis, coined by Lovell and Reiss in 1993⁷ and introduced in Spain by Novell in 1999, is used to refer to a person who is mentally retarded and has a psychiatric disorder.⁷ In the past it was not generally accepted that mentally handicapped people could, at the same time, have a mental disease. Nowadays, it is recognised that psychiatric disorders and mental retardation can occur in the same person. Consequently, these disorders can benefit from medical treatment. Dual disorder can affect up to 18%-38% of children with DS.²

The behavioural phenotype concept⁸ means "the heightened probability or likelihood that people with a given syndrome will exhibit certain behavioural and developmental sequelae relative to those without the syndrome."

In ADHD inattention is manifested as difficulty in concentrating on an activity. The person appears not to listen when they are spoken to, is disobedient, easily distracted and restless, fails to pay attention, forgets and loses things, and avoids tasks that require sustained mental effort. Hyperactivity or impulsivity are characterised by restlessness, excessive activity (running about or jumping) in inappropriate situations and these children find it difficult

Table 1 Disorders which show comorbidity with ADHD

Tourette's syndrome/Obsessive-compulsive disorder
Generalised developmental disorders
Autistic disorder
Asperger's syndrome
Generalised non-specific developmental disorder
Communication disorder (specific language disorder)
Learning difficulties
Dyslexia
Dyscalculia
Dysgraphia
Non-verbal learning disorder
Developmental coordination disorder
Behavioural disturbances
Anxiety disorders
Depression and other affective disorders

to remain seated. They talk non-stop, act without thinking and find it difficult to wait their turn. All this leads to a deterioration in their social relationships and performance at school. The symptoms in DS children may appear before the age of three.⁹

Hyperactivity and impulsivity mean that children with DS and ADHD have a high risk of hurting themselves as a result of an accident, running away, getting lost, etc.

It is necessary to establish a differential diagnosis of ADHD in children with DS. Hyperthyroidism, hearing loss and sleep apnoea¹⁰ and the side effects of certain medications, such as histamine antagonists, caffeine and adrenergic agonists must be ruled out.

ADHD does not normally appear in isolation and it is usually associated with other disorders (comorbidity)¹¹ (table 1). 40%-60% of the ADHD population has an oppositional defiant disorder, 20%-40% an antisocial behaviour disorder,¹² 25% anxiety disorders, 24% mood disorders, and 12% have a nervous tics, as well as learning difficulties. There is also a strong link between autistic spectrum disorders and ADHD and DS.¹³

Diagnosis of ADHD, assessment scales and treatment

The diagnosis of ADHD is basically clinical. It is essential to prepare a detailed medical history in order to evaluate the symptoms. It must include DSM-IV-TR clinical criteria (table 2)¹⁴ and demonstrate the existence of functional repercussions which have a significant effect on the patient's quality of life. The symptoms must be present in two or more settings, for example at school and at home. There are three ADHD subtypes: the inattentive type (if inattention signs predominate), the hyperactive-impulsive type, and the combined type (if there are manifestations of both subtypes). The most common subtype is combined ADHD and the subtype which is most difficult to diagnose is inattentive ADHD. The hyperactive-impulsive type is the least common form.

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