The neuropsychological effects of some childhood neurodevelopmental disorders

María J. García-Rubio[1]

1 Valencian International University (VIU)

Funding: No specific funding was received for this work.

Potential competing interests: No potential competing interests to declare.

Abstract

When dysfunctions occur at some stage (or several stages) in the development of the nervous system, so-called **neurodevelopmental disorders of childhood** arise.

Childhood neurodevelopmental disorders lead to or coexist with other medical, emotional and, importantly, neuropsychological conditions. This post will focus on describing the **neuropsychological consequences** of some childhood neurodevelopmental disorders.

Introduction

When dysfunctions occur at some stage (or several stages) in the development of the nervous system, so-called **neurodevelopmental disorders of childhood** arise.

Childhood neurodevelopmental disorders lead to or coexist with other medical, emotional and, importantly, neuropsychological conditions. This post will focus on describing the **neuropsychological consequences** of some childhood neurodevelopmental disorders.

1. Definition of neurodevelopmental disorders in children

Childhood neurodevelopmental disorders are varied and are defined especially on the basis of different **precipitating factors** such as: a) cause; b) age of onset and/or detection; and c) symptomatology. There are also other key points in the definition of childhood neurodevelopmental disorders such as the age of the mother, possible problems during pregnancy, genetics, and the perinatal and postnatal environment.

The **classification** on which several authors agree involves: **a) specific childhood neurodevelopmental disorders** (those classified according to DSM-V as ADHD, communication disorders, ASD, etc.), **b) early-onset childhood neurodevelopmental disorders** (anancephaly, hydrocephalus, etc.); **c) childhood neurodevelopmental disorders due to genetic variations** (e.g., Down syndrome, Williams syndrome, Angelman syndrome); d) childhood neurodevelopmental disorders due to environmental alterations (e.g., fetal alcohol syndrome, valproic acid embryopathy), Down syndrome, Williams syndrome, Angelman syndrome); **d) childhood neurodevelopmental disorders due to environmental disturbances** (e.g. foetal alcohol syndrome, valproic acid embryopathy).

There are many studies devoted to describing the neuropsychological effects derived from the neurodevelopmental disorders of childhood of the first group due to their close relationship with cognition; however, the literature on the neuropsychology of the remaining neurodevelopmental disorders is not so abundant despite the need for neuropsychological professionals to know this symptomatology in order to be able to offer the patient a comprehensive treatment.

Throughout this post, at least one childhood neurodevelopmental disorder of each type will be described with a focus on the neuropsychological effects of this condition.

2. Childhood neurodevelopmental disorders covered by DSM-V

For some years now, there have been several studies on the neuropsychological effects associated with brain disorders in children, with **ASD** being one of the most studied neurodevelopmental disorders in the clinical field and also in research, especially in high-income countries such as Spain.

2.1. *ASD and its associated neuropsychological effects*

Together with ADHD, **ASD is the** most prevalent childhood neurodevelopmental disorder in Spain. It is a denomination that currently **includes other disorders, specifically autism, pervasive developmental disorder not otherwise specified and Asperger syndrome.** Although the main cause of these diagnoses is not known at the moment, the main hypotheses focus on the **genetic cause**, so ASD is subject to a high specificity as a disorder and a high individuality as a patient. That is to say, it presents a characteristic symptomatology that allows it to be easily differentiated from other

childhood neurodevelopmental disorders but, at the same time, no two ASD patients are alike since the personal development of the patient, the support (family, school, social, etc.) and the presence or not of intellectual disability are **modulating factors** in the development of ASD; and this must be taken into account in the intervention. Although each ASD patient is unique, the **diagnosis is usually made early**, between 12 and 24 months, and includes physical medical tests, genetic assessments, and the application of a neuropsychological and psychosocial assessment battery.

The **neuropsychological effects** most present in ASD are those related to **communication** including **thinking** and **language**. Some **perceptual-motor skills** and **executive functions** are also impaired. A complete neuropsychological assessment protocol must be applied (there are several specific to ASD) to ensure which cognitive abilities are altered or preserved. In relation to **intervention**, the **American Psychological Association** has proposed that only intervention protocols with a consolidated empirical basis by experienced professionals should be applied.

3. Early-onset neurodevelopmental disorders in children

These are those that **arise during the formation and development of the** foetal **nervous system.** Depending on the stage at which the condition occurs (e.g. proliferation, migration, etc.), the infantile neurodevelopmental disorder and its associated consequences.

3.1. *Hydrocephalus and its associated neuropsychological effects*

Hydrocephalus is a neurodevelopmental disorder of childhood that usually occurs **before or around birth**. Specifically, it is a **disproportionate accumulation of cerebrospinal fluid (CSF)** inside the brain, especially in the ventricles and also in the subarachnoid space between the meninges and other structures. This excess CSF affects the dimensions of the head and brain development. Among the **most frequent causes** are the presence of a brain tumour that leads to movement of the structures and hinders optimal drainage of CSF, cerebral haemorrhage during foetal neurodevelopment and, in many cases, it is due to a congenital defect. As it is an early-onset neurodevelopmental disorder, **diagnosis** is mainly based on the application of **neurological examination techniques** (CT scan, ultrasound and MRI) that estimate the amount of extra CSF and the most affected parts of the brain. The **primary intervention** requires neurosurgery as a drain has to be inserted at brain level in order to evacuate the extra fluid.

The **neuropsychological effects** of hydrocephalus **depend on the degree of brain compression** that has taken place due to the excess CSF. Some studies have shown that these patients present alterations in **perceptual-motor skills**, with **problems in visual function**, specifically in **constructive** vision; they also suffer from **alterations in working memory**, which requires the **control** and **inhibition of information**, and some of the **executive functions** such as planning are also abnormal. The use of **pragmatic language** is also altered with this diagnosis. Therefore, the **neuropsychologist** should work on these cognitive functions with the patient after recovery from neurosurgery to ensure that the new brain configuration without the extra CSF allows these skills to develop as optimally as possible.

4. Childhood neurodevelopmental disorders due to genetic variations

Genes shape the biological characteristics of the human being, including **both** the **biological and the cognitive part**. In terms of brain development, genetics also plays a key role in guiding the evolution of the brain, not only for the species but also for each individual within the species. When genetics presents a certain anomaly during brain development, the so-called **neurodevelopmental disorders in children** arise **due to** genetic **variation**.

4.1. *Williams syndrome and its associated neuropsychological effects*

It is a rare genetic disorder, compared to Down's Syndrome, for example, but it has a high specificity and affects several of the fundamental cognitive functions, which is why it was selected for this post. This neurodevelopmental disorder in children is caused by a **microdeletion of chromosome number 7,** which produces **pathologies of different physical systems** (cardiac malformations, metabolic problems related to different substances), and **phenotypic traits** such as a pixie face.

Diagnosis is early and they are usually treated from an early age with neurological, physical and behavioural screening tests. Early detection of this neurodevelopmental disorder in children is essential in these patients as numerous research studies have shown that **early intervention can maintain certain cognitive abilities.**

With regard to **neuropsychological waiting**, professionals in this field must be familiar with the disorder and establish an individualised cognitive profile since, although they are associated with **moderate to severe mental retardation**, some skills such as **language and memory are preserved**, for example, they recognise faces easily. It has also been shown that these patients are **very attracted to music**, being sensitive to sounds, and tend to establish **affective bonds** with those around them. However, they have **dysfunction in visuospatial and/or visuoperceptive skills**. Therefore, it is recommended that the neuropsychological professional should apply a neuropsychological assessment battery focusing on perceptual skills (visual, auditory, sensory, etc.) in order to establish the patient's strengths and weaknesses from a neuropsychological point of view and thus be able to draw up an intervention plan.

5. Childhood neurodevelopmental disorders due to environmental variations

When **neurodevelopmental disorders in children** are a consequence of **factors present in the environment**, a causal relationship is hypothesised, although this cannot be proven in all cases. The nervous system begins its development in the first weeks of gestation (at approximately 21 days) and progresses through different phases, some being more sensitive than others. It has been shown that **exposure of the nervous system** to certain **toxic or pathogenic agents** during some of these phases can lead to neurodevelopmental disorders such as fetal alcohol syndrome, which will be described below. The **source of toxicity** does not always come from the pregnant mother (alcohol, tobacco, mental health problems, etc.) but may also be present in the environment in which she lives during gestation, such as exposure to xenobiotic substances from industrial, agricultural or urban activity in the surroundings where she lives, for example.

5.1. *Fetal Alcohol Syndrome and its associated neuropsychological effects*

This syndrome is included within the **Fetal Alcohol Spectrum Disorder (FASD)** which implies a broader concept that groups together **all possible diagnoses** with symptoms of a physical, mental, cognitive, behavioural and emotional nature suffered by a **child exposed to alcohol during prenatal development.**

Fetal Alcohol Syndrome (FAS) is the **ultimate expression** of infantile neurodevelopmental disorder within **FASD**. The **cause** is well known as alcohol exposure during prenatal life is the main cause of malformations during nervous system development, as we saw in Table 3. Among the **symptoms associated** with the diagnosis of FAS are **systemic** and especially **cerebral growth retardation** leading to alterations in synaptic levels and structure formation; therefore, **craniofacial malformations** are very characteristic of these patients, leading to significant **cognitive and behavioural problems**. Some patients also have **problems with height and weight** and episodes of **clinical seizures** are also frequent.

At the **neuropsychological level**, specific cognitive areas are affected, with **learning problems** being very frequent in these patients. The most frequent studies highlight **severe problems in processing**, **verbal comprehension**, **perceptual reasoning** and **visuospatial reasoning**, together with behavioural disorders with defiant overtones and transgression of rules and limits. Although more and more is known about the **neurocognitive profile** of FAS, a specific profile for this syndrome has not yet been established as some dysfunctions are masked by other emotional disorders produced by the lack of attachment or the feeling of abandonment that most of these patients suffer from an early age. In this case, the **neuropsychologist** should apply a standard neuropsychological assessment battery and work on deficits and strengths with the patient and family.

6. Conclusions

Although there is no stable classification to break down **childhood neurodevelopmental disorders**, throughout this entry some of these disorders have been described according to their aetiology, stage of onset, and comorbid consequences. Specifically, a disorder within each type has been described with special emphasis on the **neuropsychological effects** they entail so that **neuropsychological professionals** have more information about them and can carry out the work ahead not only in neurodevelopmental disorders included in the DSM-V that are better known but especially in other disorders arising from a genetic or environmental malformation or those of early onset to which attention is given from other health fields (neurosurgery, neuroenfirmary, etc.), neuroenfirmary, etc.) without paying too much attention to the neuropsychological effects that these patients present.

References

- Betts, J., Dawe, S., Eggins, E., Shelton, D., Till, H., Harnett, P., & Chandler-Mather, N. (2019). PROTOCOL: Interventions for improving executive functions in children with Fetal Alcohol Spectrum Disorder: Systematic review and

meta-analysis. *Campbell Systematic Reviews*, *15*(1-2), e1009.

- Braconnier, M. L., & Siper, P. M. (2021). Neuropsychological assessment in autism spectrum disorder. *Current Psychiatry Reports*, *23*(10), 1-9.
- De la Torre, G. G., Martin, A., Cervantes, E., Guil, R., & Mestre, J. M. (2017). Attention lapses in children with spina bifida and hydrocephalus and children with attention-deficit/hyperactivity disorder. *Journal of clinical and experimental neuropsychology*, *39*(6), 563-573.
- García, R. J. G., & Agramonte, M. D. L. Á. R. (2020). Comorbidity in patients with neurodevelopmental disorders. *Cuban Journal of Pediatrics*, *92*(4).
- Gonzalvo, G. O. (2011). Frequency of fetal alcohol syndrome in institutionalized children from Eastern European countries. *Journal of neurology*, *53*(2), 127-128.
- Klein-Tasman, B. P., van der Fluit, F., & Mervis, C. B. (2018). Autism spectrum symptomatology in children with Williams syndrome who have phrase speech or fluent language. *Journal of autism and developmental disorders*, *48*(9), 3037-3050.
- Lange, S., Shield, K., Rehm, J., Anagnostou, E., & Popova, S. (2019). Fetal alcohol spectrum disorder: Neurodevelopmentally and behaviorally indistinguishable from other neurodevelopmental disorders. *BMC psychiatry*, *19*(1), 1-10.
- Lense, M. D., Ladányi, E., Rabinowitch, T. C., Trainor, L., & Gordon, R. (2021). Rhythm and timing as vulnerabilities in neurodevelopmental disorders. *Philosophical Transactions of the Royal Society B*, *376*(1835), 20200327.
- Livingston, L. A., & Happé, F. (2017). Conceptualising compensation in neurodevelopmental disorders: Reflections from autism spectrum disorder. *Neuroscience & Biobehavioral Reviews*, *80*, 729-742.
- Mattson, S. N., Crocker, N., & Nguyen, T. T. (2011). Fetal alcohol spectrum disorders: neuropsychological and behavioral features. *Neuropsychology review*, *21*(2), 81-101.
- Mikkelsen, R., Rødevand, L. N., Wiig, U. S., Zahl, S. M., Berntsen, T., Skarbø, A. B.,... & Wester, K. (2017). Neurocognitive and psychosocial function in children with benign external hydrocephalus (BEH)-a long-term follow-up study. *Child's Nervous System*, *33*(1), 91-99.
- Parenti, I., Rabaneda, L. G., Schoen, H., & Novarino, G. (2020). Neurodevelopmental disorders: from genetics to functional pathways. *Trends in Neurosciences*, *43*(8), 608-621.
- Riley, E. P., Infante, M. A., & Warren, K. R. (2011). Fetal alcohol spectrum disorders: an overview. *Neuropsychology review*, *21*(2), 73.
- Serrano-Juárez, C. A., Prieto-Corona, D. M. B., & Yáñez-Téllez, M. G. (2018). Neuropsychological intervention in a case of a girl with Williams Syndrome. *Cuadernos de Neuropsicología/Panamerican Journal of Neuropsychology*, *12*(2).
- Vacas, J., Antolí, A., Sánchez-Raya, A., & Cuadrado, F. (2020). Analysis of Cognitive Profiles in Child Clinical Population with Neurodevelopmental Disorders. *Revista Iberoamericana de Diagnóstico y Evaluación-e Avaliação Psicológica*, *1*(54), 35-46.
- Vivanti, G., Hamner, T., & Lee, N. R. (2018). Neurodevelopmental disorders affecting sociability: recent research advances and future directions in autism spectrum disorder and Williams syndrome. *Current neurology and*

neuroscience reports, *18*(12), 1-9.

- Zielińska, D., Rajtar-Zembaty, A., & Starowicz-Filip, A. (2017). Cognitive disorders in children's hydrocephalus. *Neurologia i neurochirurgia polska*, *51*(3), 234-239.
- Zwick, G. P. (2017). Neuropsychological assessment in autism spectrum disorder and related conditions.*Dialogues in clinical neuroscience*, *19*(4), 373.