

## *Neuropsychological assessment in autism spectrum disorder and related conditions*

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*Neuropsychological assessment provides a profound analysis of cognitive functioning in people with autism spectrum disorder (ASD). Individuals on the autistic spectrum often show a high level of anxiety and are frequently affected by comorbidities that influence their quality of life. Yet, they also have cognitive strengths that should be identified in order to develop effective support strategies. This article presents an overview of five cognitive areas that are essential for neuropsychological evaluation (ie, intelligence, attention, executive function, social cognition, and praxis) and explores the underlying causes of behavioral problems in persons with ASD. Furthermore, it stresses the importance of meticulous neuropsychological testing with regard to cognitive remediation, a method that can help to enhance single cognitive processes in a targeted manner. Objective test results suggest it might be possible to promote an improved sense of coherence. In line with the salutogenic model, this may be fundamental for human health and well-being.*

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*Dialogues Clin Neurosci.* 2017;19:373-379.

### Introduction

Since the beginning of the 20th century,<sup>1</sup> the concept of autism has undergone various modifications, as reflected in current diagnostic tools. Whereas the tenth version of the *International Statistical Classification of Diseases and Related Health Problems (ICD-10)*<sup>2</sup> still uses a categorical approach in which autism is listed as one of eight pervasive developmental disorders, the fifth edition of the *Diagnostic and Statistical Manual of Mental Disorders (DSM-5)*<sup>3</sup> offers a more dimensional description of these conditions by considering them as a continuum. As a result, the *DSM-5* uses the term “autism spectrum disorder” (ASD), which is also anticipated to appear in the upcoming *ICD-11*. For a simplified summary, these differences are illustrated in *Table I*.

According to the *DSM-5*, ASD is characterized by “...persistent deficits in social communication and social interaction across multiple contexts...,” as well as “...restricted, repetitive patterns of behavior, interests, or activities...”<sup>3</sup>

Beyond all nosology, De Clercq<sup>4</sup> pleads for an understanding of autism from within and points out that

**Keywords:** attention; autistic spectrum; executive function; intelligence; neuropsychological assessment; praxis; salutogenesis; social cognition

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the testimony of high-functioning people with autism is a precious source of information that may help to improve the understanding of the autistic spectrum.

Tammet,<sup>5</sup> for example, provides deep insight into his life with Asperger syndrome in his memoir *Born on a Blue Day*, in which he describes his fears and joys as well as his special interest for numbers and languages. He also mentions his preference for detailed thinking, which appears to be a frequent characteristic of persons with ASD.<sup>5,6</sup> In addition, Schovanec<sup>7</sup> illustrates that the human personality is far too complex to be reduced to one aspect, refusing thereby to define himself solely by autism. In a humorous way, he raises philosophical questions about autism and normality by explaining that he does not feel autistic at all when he is alone, but encounters various difficulties in social interactions.

In fact, people with ASD often show a high level of anxiety and are frequently affected by comorbidities,<sup>3</sup> which influence their quality of life. Therefore, it is crucial to analyze their cognitive and emotional functioning and to adapt their social and material environment to their needs. A priori, neuropsychological assessment is predestined for this purpose. However, due to the restricted interests of individuals on the autistic spectrum, their motivation might strongly fluctuate throughout the testing process, leading to a performance that does not necessarily reflect their cognitive potential. Thus, it is essential to explicitly point out the importance of the examination, whenever the first signs of disinterest appear.

ICD-10	DSM-5
Childhood autism (F84.0)	Autism Spectrum Disorder (299.00)
Atypical autism (F84.1)	
Rett syndrome (F84.2)	
Other childhood disintegrative disorder (F84.3)	
Overactive disorder associated with mental retardation and stereotyped movements (F84.4)	
Asperger syndrome (F84.5)	
Other pervasive developmental disorders (F84.8)	
Pervasive developmental disorder, unspecified (F84.9)	

**Table 1.** Pervasive developmental disorders (ICD-10) vs autism spectrum disorder (DSM-5), excluding Rett syndrome. *DSM-5*, *Diagnostic and Statistical Manual of Mental Disorders*, 5th edition<sup>3</sup>; *ICD-10*, *International Statistical Classification of Diseases and Related Health Problems*, 10th revision.<sup>2</sup>

## Neuropsychological assessment

The aim of neuropsychological assessment is to identify cognitive strengths and weaknesses, to assess their consequences in everyday life, and to organize appropriate support. Moreover, a thorough evaluation may be helpful for refining the patient's diagnosis. Typically, neuropsychological assessment is performed based on the patient's anamnesis and complaints as well as clinical observations. Because certain tests may have to be modified without compromising the validity of the results, people with ASD should be examined by well-trained, experienced neuropsychologists. Above all, the investigation should highlight the uniqueness of each person with ASD, regardless of the severity of the disorder. In the following sections, five essential areas of a comprehensive neuropsychological evaluation are presented more closely (ie, intelligence, attention, executive function, social cognition, and praxis).

## Intelligence

The assessment of intelligence is fundamental for any further neuropsychological evaluation and provides indications for many well-founded interventions. The Wechsler Intelligence Scales are the most frequently used intelligence tests in the world. They are available in several versions, each corresponding to a certain age range. The fourth edition of the Wechsler Adult Intelligence Scale (WAIS-IV)<sup>8</sup> may serve as an example for illustrating some basic principles. In short, this psychometric test distinguishes between linguistic and practical intelligence, which allows for a differentiated estimation of the patient's level of intelligence. It includes four primary indexes, the Verbal Comprehension Index (VCI), the Perceptual Reasoning Index (PRI), the Working Memory Index (WMI), and the Processing Speed Index (PSI). These indexes, consisting of ten core and five supplemental subtests, form the so-called cognitive profile, which is either homogeneous or heterogeneous. Homogeneity is given when there are no significant differences between the index scores. This is also a condition sine qua non for the determination of the overall intelligence quotient (IQ), which is only meaningful and representative of the patient's cognitive abilities when the gap between the primary indexes is not too high. Thus, from a clinical point of view, intellectual disability, a frequent co-

occurrence of ASD,<sup>3</sup> should not be diagnosed in the case of heterogeneous profiles.

There is also some evidence that the Wechsler Intelligence Scales might underestimate the intelligence of people with autism.<sup>9,10</sup> Hence, the test results must always be interpreted with great caution to avoid stigmatizing and misleading assumptions. In addition, it may be pointed out that the Wechsler Nonverbal Scale of Ability (WNV)<sup>11</sup> is often the more adequate choice for persons with low-functioning ASD, since it can be carried out without speaking.

Another issue of significant relevance is the relationship between cognitive profiles and ASD symptoms. In the research literature, there are many conflicting findings. Some authors, for example, associate autistic features with discrepancies between verbal and nonverbal abilities,<sup>12</sup> whereas others relativize this assumption.<sup>13</sup> However, considering the conceptual changes of autism, it makes little sense to compare current studies with data collected more than a decade ago.

Recent findings emphasize sex-related differences in the cognitive profiles of persons with ASD, which is important for both diagnosis and adequate support measures. As reported by Lehnhardt et al.,<sup>14</sup> males with ASD show higher verbal abilities, whereas females with ASD demonstrate higher processing speeds and better executive functions. The female strengths might be related to social skills, allowing women to mask their autistic symptoms in a more effective way than men.

Oliveras-Rentas et al.<sup>15</sup> also underline the importance of processing speed. According to their results, processing speed task performance correlates positively with communication abilities as well as negatively with communication symptoms. Nevertheless, they note that the subtests of the PSI do not clearly distinguish between cognitive processing speed and motor speed, so further research is required to clarify this matter.

Altogether, these studies demonstrate that the analysis of the cognitive profile is by far more important than the measurement of the overall IQ. Basically, the patient's performance should always be interpreted on a subtest level in conjunction with his/her respective living conditions. Moreover, support measures must be individually tailored and oriented toward the patient's needs. Consequently, an even more comprehensive evaluation of individual cognitive functions and their underlying processes may be required to effectively reduce stressful situations for people with ASD.

## Attention

It is well-established that attention governs the proper functioning of other cognitive functions and that even minor disturbances can have far-reaching consequences in everyday life.<sup>16</sup> Actually, there are many theoretical models of attention that illustrate the complexity of this function, which results from the activity of widely distributed neuronal networks in the human brain.<sup>17</sup> According to the *DSM-5*,<sup>3</sup> individuals on the autistic spectrum often present with attention abnormalities. Since the *DSM-5* no longer considers ASD as an exclusion criterion for attention-deficit/hyperactivity disorder (ADHD), there is growing evidence that both disorders often co-occur.<sup>18</sup> Moreover, recent findings suggest that this combination might weaken the patient's adaptive skills far more than each one of these disorders alone. Therefore, individuals affected by both disorders might not be able to improve their adaptive functioning in an effective way if experts merely focus on ASD symptoms.<sup>19</sup>

In line with neuropsychological observations, it may be added that attention deficits often cause fatigue and slow down the patient's cognitive performance. Since the WMI and the PSI of the Wechsler Intelligence Scales are strongly dependent on attention, their relative weakness compared with the other primary indexes could already indicate attention difficulties. Nevertheless, both slight fluctuations of attention and ADHD must always be confirmed by a meticulous examination.

There are many neuropsychological assessment tools that can be used to explore various facets of attention. Depending on the equipment, either computerized tests or paper-pencil tests are used. Testing is always guided by clinical observations but should at least cover the patient's *alertness* as well as his/her *sustained*, *divided*, and *selective attention*.

*Alertness* can be defined as "...the ability to temporarily increase and sustain the intensity of attention."<sup>20</sup> If this attention process is deficient, patients may have serious difficulties reacting to certain hazardous situations in a timely manner (eg, braking to avoid an accident).

*Sustained attention* "...involves focusing attention on a mentally demanding activity for a sustained period of time."<sup>21</sup> This is obviously a prerequisite for any kind of efficient activity (eg, maintaining concentration at work).

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*Divided attention* is, as the term suggests, "...the ability to perform different tasks simultaneously."<sup>20</sup> If this process does not function properly, patients might also experience a variety of difficulties (eg, listening to the teacher's voice while writing down information; holding a conversation with several people).

*Selective attention* describes "...the ability to respond quickly to relevant stimuli and to suppress inappropriate responses."<sup>20</sup> As reported by Stevens and Bavelier,<sup>22</sup> this function of attention particularly influences language, literacy, and math skills. Therefore, it is related, among many other factors, to academic achievement. It may be noted that neuropsychological tests distinguish between visual and auditory modality, which is quite important for pedagogical support measures.

For pragmatic reasons, other attention processes can be disregarded (eg, *vigilance*, *focused attention*, *visuospatial attention*). However, since persons with ASD might not spontaneously talk about their attention difficulties, it is important to consult their relatives and to estimate the severity of the symptoms in everyday life. If necessary, ADHD diagnostic criteria can also be evaluated at this point. According to the *DSM-5*,<sup>3</sup> symptoms must occur in at least two different settings. In order to achieve a broad overview of the patient's behavior, structured questionnaires can be helpful. The third edition of the *Conners*,<sup>23</sup> for example, has received a *DSM-5* update and provides teacher, parent, and student perspectives with regard to ADHD core symptoms and associated problems.

To meet the quality standards of clinical neuropsychology, a reliable ADHD diagnosis should always take into account the patient's cognitive and behavioral profile and be based on a careful assessment of all central attention processes. In fact, such a detailed analysis is extremely important with regard to cognitive remediation and can help to improve specific functions of attention in a targeted manner.

## Executive function

Unfortunately, there are many coexisting definitions of executive function (EF)<sup>24</sup> that jeopardize the clarity of this concept. However, it is widely acknowledged that EF is an umbrella term used for multiple cognitive functions,<sup>25</sup> allowing for successful adaptation to complex environmental conditions. A recent meta-analysis emphasizes a broad executive dysfunction in people with

ASD,<sup>26</sup> even though these impairments are not specific to ASD.<sup>27</sup> According to research, EF deficits significantly lower the quality of life in individuals with ASD,<sup>28</sup> which underlines the importance of assessing these functions.

Although isolated executive weaknesses do not necessarily lead to disability if the environment provides adequate compensations, clinical neuropsychologists should at least examine the patient's *inhibition*, *working memory*, *updating*, *planning*, and *cognitive flexibility*. In this context, it may be pointed out that formal tests possibly lack ecological validity.<sup>29,30</sup> Therefore, experts should also rely on clinical observations in real life.

*Inhibition* is often associated with behavioral control and can be summarized, in simplified terms, as "...the ability to suppress unwanted reactions."<sup>20</sup> If this function is deficient, persons with ASD may experience various difficulties, especially in social interactions (eg, inhibition of socially inappropriate stress reactions).

In accordance with Baddeley,<sup>31</sup> *working memory* provides a temporary storage and manipulation of information that can be linked with long-term memories. Thus, it significantly influences learning, reasoning, and problem-solving skills but is also required in social and professional contexts (eg, having a conversation without forgetting important contents; performing multiple instructions). In fact, *working memory* impairments can affect the personal development of individuals with ASD in almost every area of life.

*Updating* can be defined as "...the ability to renew memory contents in a controlled and goal-directed manner."<sup>20</sup> If this function does not work properly, patients can be extremely unbalanced in terms of thinking (ie, rumination) and social interaction (eg, comprehension difficulties).

*Planning* refers to "...a complex, dynamic operation in which a sequence of planned actions must be constantly monitored, re-evaluated, and updated."<sup>32</sup> As a result, successful *planning* is inconceivable without the assistance of other executive functions (eg, *working memory* and *updating*), which highlights their interconnectivity. However, *planning* deficits can easily lead to specific problems in everyday life that require specific support measures (eg, cooking a meal, organizing leisure activities, running a household).

*Cognitive flexibility* is "...the ability to shift to a different thought or action according to changes in a situation."<sup>32</sup> Recent findings suggest that *cognitive flexibility* problems might be associated with anxiety-related



symptoms in persons with ASD.<sup>33</sup> Overall, this specific deficit makes it difficult to adapt to constantly changing environmental conditions and thereby increases the stress level of individuals on the autistic spectrum.

A priori, there are even links between EF and social cognition;<sup>34,35</sup> this underlines the complexity of these cognitive functions.

### Social cognition

Social cognition refers to a range of cognitive processes that allow people to interact and to understand each other. A variety of studies report social cognition impairments in individuals with ASD.<sup>36-40</sup> Therefore, neuropsychological assessment should examine all core components in this field, namely *emotion perception, theory of mind, attributional style, and social perception and social knowledge*. Since there are only a few standardized neuropsychological tests available, the evaluation is partly based on qualitative observations.

*Emotion perception* implies the ability to recognize and to identify emotions in other people, which is crucial for many kinds of social interaction. In fact, if patients do not accurately perceive emotional information, they run the risk of misinterpreting the tone of the voice, the body language, or the facial expressions of their interlocutors, which might have negative consequences on their social adjustment.<sup>41</sup>

*Theory of mind* refers to the ability to reflect on the contents of one's own and others' minds.<sup>42</sup> If persons with ASD are not able to imagine other people's thoughts, beliefs, desires, or intentions by ignoring their own point of view, they might misunderstand many social interactions, leading to social isolation in the long term.

*Attributional style* describes how people explain the causes of positive and negative events in their life. Usually, events have a multifactorial origin. Yet, if the *attributional style* is impaired, patients may generate exaggerated explanations, attributing the responsibility of an event to a single cause. For example, some patients are firmly convinced that all positive events happen thanks to other people or by chance. In return, they always blame themselves for any negative event (depressive bias).<sup>43</sup> Other patients suspect fellow human beings to have hostile intentions, even when their behavior is completely harmless (hostile bias).<sup>44</sup>

Actually, there are many biases in causal attributions that might complicate the patient's life, especially

if they are very pronounced and thus pathological. Although some biases tend to be related to diseases, such as schizophrenia,<sup>45,46</sup> it is important to assess the *attributional style* of persons with ASD comprehensively, with regard to differential diagnosis and comorbidities.

*Social perception and social knowledge* allow people to identify and to understand social roles, rules, and common principles in social situations.<sup>47</sup> If these processes are deficient, people may have difficulties behaving according to social norms, which disadvantages them in many ways.

Obviously, all these components of social cognition are essential for any socially appropriate behavior. Once again, strengths and weaknesses should be assessed in conjunction with the individual living conditions of the patient in order to organize effective support measures.

### Praxis

Praxis refers to the ability to conceptualize, plan, and successfully carry out purposeful motor actions.<sup>48</sup> It is widely acknowledged that ASD is associated with abnormalities in motor skills,<sup>3</sup> ranging from clumsiness to dyspraxia,<sup>49</sup> which is also called developmental coordination disorder (DCD).<sup>50</sup>

Praxis problems may have far-reaching consequences, both in the private (eg, dressing, washing, using utensils) and public spheres (eg, failure at school or at work, due to organizational difficulties, fatigue, and slowness). But there is also growing evidence that praxis influences social skills, which is even more important with regard to autism spectrum features (ie, impairments in social communication and social interaction).

For example, Cassidy et al<sup>51</sup> emphasize that motor coordination skills are crucial for empathy and effective social skills. Other findings also suggest that praxis impairments negatively affect social activities in children with ASD, such as age-related play and leisure activities.<sup>52</sup>

Consequently, a conscientious neuropsychological assessment of persons with ASD cannot disregard the evaluation of their praxis skills and should appraise all major subtypes of DCD<sup>53</sup> if clinical observations indicate such disturbances.

### Conclusion

Neuropsychological assessment allows for a profound analysis of the cognitive functioning of individuals on the autistic spectrum, even though the ecological valid-

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ity of certain tests needs to be improved. Moreover, the evaluation is not very time-consuming, since it takes no longer than approximately 6 hours to assess all five core areas presented in this article. Yet, 5 additional hours are required to analyze the data and to prepare a detailed report including all test results, as well as specific recommendations, such as individually tailored support strategies and cognitive remediation.<sup>20,54,55</sup>

With the patient's consent, these neuropsychological findings may be shared with relatives and experts of the medico-psychosocial field. Since behavioral problems represent merely the tip of the iceberg,<sup>56,57</sup> all stakeholders should focus on the underlying causes of these symptoms to effectively prevent stress reactions in people with ASD. In this context, clinical neuropsychologists may offer many well-founded working hypotheses.

Based on objective test results and in line with salutogenesis, it is also possible to promote the patient's sense of coherence, which appears to be fundamental for human health and well-being. Therefore, all stake-

holders should ensure that the patient's daily activities are as comprehensible, manageable, and meaningful as possible. According to Antonovsky,<sup>58</sup> comprehensibility implies that internal and external stimuli are perceived as orderly, consistent, structured, and clear, making future events predictable or at least explainable. Manageability refers to the extent to which individuals perceive that they have appropriate resources to master complex demands and requirements. Meaningfulness signifies that challenges are considered worthwhile, especially in an emotional sense.

Finally, from a clinical point of view, the term autism spectrum disorder seems to be poorly chosen, since it does not sufficiently reflect areas of cognitive strengths in individuals on the autistic spectrum. Therefore, neuropsychological assessment should emphasize these strengths all the more. □

**Acknowledgments/Conflict of Interest:** I would like to thank Katharina and Magnus Benedict for their proofreading of my article. The author declares that he has no relevant or material financial interests that relate to the content described in this article.

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### **La evaluación neuropsicológica en el trastorno del espectro autista y los cuadros relacionados**

*La evaluación neuropsicológica facilita un profundo análisis del funcionamiento cognitivo en las personas con trastorno del espectro autista (TEA). Los sujetos con el espectro autista a menudo muestran un alto nivel de ansiedad y presentan con frecuencia comorbilidades que influyen en su calidad de vida. Sin embargo, ellos también tienen fortalezas cognitivas que deben ser identificadas para desarrollar estrategias de soporte efectivas. Este artículo entrega una panorámica de cinco áreas cognitivas que son esenciales para la evaluación neuropsicológica (como inteligencia, atención, función ejecutiva, cognición social y praxis) y explora las causas subyacentes de los problemas conductuales en personas con TEA. Además, se enfatiza la importancia de una evaluación neuropsicológica rigurosa en relación con la remediación cognitiva, un método que puede ayudar a mejorar los procesos cognitivos individuales de una manera específica. Los resultados de las pruebas objetivas sugieren que podría ser posible fomentar un mejor sentido de coherencia en las personas con TEA. En línea con el modelo salutífero, esto puede ser fundamental para la salud y el bienestar humanos.*

### **Évaluation neuropsychologique du trouble du spectre de l'autisme et des maladies qui y sont liées**

*L'évaluation neuropsychologique permet une analyse approfondie du fonctionnement cognitif chez les sujets atteints de trouble du spectre de l'autisme (TSA). Ces derniers présentent souvent une anxiété importante et souffrent fréquemment de comorbidités influant sur leur qualité de vie. Ils ont pourtant également des aptitudes cognitives qui devraient être identifiées afin de développer des stratégies de soutien efficaces. Cet article présente une vue d'ensemble des cinq domaines cognitifs essentiels pour l'évaluation neuropsychologique (c'est-à-dire, l'intelligence, l'attention, la fonction exécutive, la cognition et la pratique sociales) et explore les causes sous-jacentes des problèmes comportementaux chez les sujets atteints de TSA. De plus, l'importance d'une évaluation neuropsychologique rigoureuse y est soulignée en ce qui concerne la remédiation cognitive, une méthode qui peut améliorer de façon ciblée des processus cognitifs particuliers. D'après des résultats objectifs, le sens de la cohérence pourrait être amélioré chez les sujets ayant un TSA. Ceci pourrait s'avérer crucial pour la santé et le bien-être chez l'homme dans le cadre d'une démarche basée sur la salutogenèse (promotion de la santé).*