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Introduction: Children with Noonan syndrome, the most prevalent RASopathy, exhibit a characteristic neurocognitive phenotype. Specific difficulties in learning, memory, and executive function are among the most frequently observed manifestations documented in the scientific literature.
Objectives: The present study aimed to measure specific learning difficulties and intelligence in children with RASopathies.
Methods: A total sample of 47 patients (55.3% male) aged 3–16 years (M = 8.6, $S.D.$ = 3.1) was recruited from various Spanish regions (Murcia, Alicante, Gandía, Valencia, Castellón de la Plana, Tarragona, Cantabria, Asturias, Zaragoza, Madrid, Badajoz, Huelva, Sevilla and Cádiz), within the framework of The Grey Matter project. Most patients were diagnosed with Noonan syndrome, and some had Cardiofaciocutaneous syndrome. The most frequent mutations were PTPN11 (72.3%), RIT1 (6.4%), and unknown mutations in 12.8% of patients. Patients were assessed using the Spanish version of the Wechsler Preschool and Primary Scale of Intelligence, Fourth Edition (WPPSI-IV) or the Wechsler Intelligence Scale for Children, Fifth Edition (WISC-V) according to age; the Spanish Reading Processes Evaluation Battery (PROLEC) according to the appropriate age range (from 6 years); and the Spanish Writing Processes Evaluation Battery (PROESC), from 8 years.
Results: On the one hand, 32.3% presented with severe difficulty in reading words. In turn, 30% of the children showed severe difficulty in reading comprehension of texts when they were read by an examiner, and 29% after reading by themselves. On the other hand, 56% of the children presented severe orthographic difficulty in writing words and 50% presented severe difficulty in written expressions. Finally, regarding intelligence, the mean total IQ of the children was 84.7 points ($S.D.$ = 15.1), with better scores on the Verbal Comprehension Index (M = 89.4; $S.D.$ = 16), Visual Spatial Index (M = 87.7; $S.D.$ = 13.8), and Fluid Reasoning Index (M = 88.6; $S.D.$ = 11.9), in contrast with the Working Memory Index (M = 84.3; $S.D.$ = 13.2), and the Processing Speed Index (M = 83.2; $S.D.$ = 14.4). In turn, 42.6% of children had low global intelligence scores (IQ ≤ 79).
Conclusions: In conclusion, children with RASopathies, particularly those diagnosed with Noonan syndrome, need educational support to compensate for all these significant academic difficulties.

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Everyday Executive Functioning in Paediatric Patients Awaiting Solid Organ or Haematopoietic Stem Cell Transplantation

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Introduction: Paediatric patients with severe diseases awaiting solid organ or haematopoietic stem cell transplantation are known to experience cognitive impairments, particularly in executive functioning. Neuropsychological pre-transplant evaluation serves as a baseline for identifying executive functioning deficits that may affect the medical and psychosocial aspects of care. This is clinically relevant because patients with poorer executive functioning may show decreased adherence to medical treatments, face greater challenges in coping with their illness, and be more likely to require educational adaptations, including curricular or methodological adjustments based on pedagogical criteria.
Objectives: This study aimed to assess everyday executive functioning in paediatric patients awaiting transplantation using an ecologically valid measure.
Methods: A total of 49 patients (59.2% male) aged between 6 and 18 years (M = 11.4, $S.D.$ = 3.5) were recruited from La Paz University Hospital (Madrid, Spain). Patients were awaiting for various types of organ transplants (kidney: 26, heart: 4, lung: 4, hepato-renal: 3, or liver: 2 patients) or haematopoietic stem cells (10 patients). Patients were assessed using the Spanish parent-reported version of the *Behavior Rating Inventory of Executive Function, Second Edition* (BRIEF-2) with age- and sex-adjusted norms from the Spanish general population. The three BRIEF-2 composite scores were analysed (the Cognitive, Emotional and Behavioral Regulation Indexes), and clinically relevant scores were set at $T \geq 65$ points.
Results: Clinically significant levels of executive deficits were observed in 33.3% of patients regarding cognitive regulation problems, in 32.7% regarding emotional regulation difficulties and in 26.5% regarding behavioural regulation problems.
Conclusions: Between 1 in 4 and 1 in 3 patients have shown some type of executive regulation difficulties. The early identification of executive functioning deficits in paediatric transplant candidates is crucial. Incorporating standardised ecologically valid measures, such as the BRIEF-2, into routine assessments can help detect neuropsychological impairments at an early stage, allowing timely therapeutic or preventive interventions (e.g., psychological prehabilitation). This approach can improve medical outcomes and quality of life and guide educational adaptations to support academic performance.

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