

MALATTIE EMORRAGICHE CONGENITE E ACQUISITE

COGNITIVE, ADAPTIVE ABILITIES AND BEHAVIORAL SYMPTOMS IN CHILDREN AND ADOLESCENTS WITH HEMOPHILIA: A PILOT STUDY.

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Background and Aims:

Hemophilia, a complex chronic illness, poses risks to children's cognitive, behavioral, and adaptive development. Its daily management can also increase parental stress and the likelihood of dysfunctional parenting. This pilot study aims to assess cognitive, behavioral, and adaptive functioning in children and adolescents with hemophilia, to identify early risk indicators and guide prevention and support strategies.

Methods:

Twelve boys aged 6-14 years (M=9.6, SD=2.4) with severe hemophilia A or B, treated per current standards, were assessed using:

- Adaptive Behavior Assessment System II (Ferri et al., 2014), to evaluate adaptive skills and social adjustment capacity;
- Raven's Matrices (Raven, 2008), to assess non-verbal cognitive abilities;
- Parenting Stress Index - Short Form (Abidin, 1995), to measure parental stress related to the management of chronic illness;
- Questionnaire on Physical and Verbal Aggression, Prosocial Behavior, and Emotional Instability (Caprara et al., 1992), to assess emotional and behavioral characteristics.

The group was divided into two groups based on the presence or absence of a neurodevelopmental disorder, and the subgroups were compared across cognitive, behavioral, and adaptive functioning domains using non-parametric Wilcoxon tests.

Results:

Cognitive functioning was within the normal range

(M=106.4, SD=14.2). Adaptive functioning was in the low-average range (M=90.7, SD=20.9), with greater difficulties in the practical domain (M=88.2, SD=19.9). Parental stress was elevated (M=86.7, SD=75.0), especially in parental distress and perception of the child as difficult. Behavioral assessments showed moderate emotional instability and good prosocial behavior, with discrepancies between parent and child reports—particularly in perceived aggressiveness and emotional instability (linked to impulsivity and attention), which were rated higher by parents.

Parents reported concerns about their child's social adaptive behavior, especially regarding emotional instability (difficulty focusing, staying still, and self-regulating). A comparison between children with (n=6) and without (n=6) neurodevelopmental disorders revealed significantly lower adaptive scores in the former: global (M=77.7, SD=16.7, p=0.01), conceptual (M=81.2, SD=20.3, p=0.03), and social (M=79.2, SD=12.5) vs. those without disorders (global M=104.3, SD=15.5; conceptual M=106, SD=14.8; social M=108.5, SD=13.7). Differences in non-verbal IQ and practical skills were not significant.

Conclusions:

Children with hemophilia show preserved cognitive abilities but reduced adaptive functioning, especially in practical skills. Those with neurodevelopmental disorders are more vulnerable behaviorally and adaptively, with increased risk for ADHD and learning difficulties. Higher parental stress is associated with lower child adaptive functioning. These findings support the need for integrated care, including psychological monitoring and family support.

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