Cognitive, Emotional, and Behavioral Problems of Children with Hemophilia

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ABSTRACT

Background: Children with hemophilia are prone to a variety of psychological problems due to some limitations associated with the disease. We aimed to compare the cognitive, emotional, and behavioral problems of children with hemophilia to healthy children.

Methods: This study was performed on 65 children with hemophilia and 65 healthy individuals as the control group who were between the ages of 7 and 12 years in Children's Hospital. The Child Behavior Checklist (CBCL) was used to identify emotional/behavioral problems and Wisconsin Card Sorting Test (WCST) to evaluate cognitive problems.

Results: The results showed that children with hemophilia obtained lower scores in activity, academic performance, and overall competence variables. Children with hemophilia in comparison to healthy children showed more internalizing and externalizing problems and emotional and behavioral deficits. Also they demonstrated more impairment in executive functions than healthy children.

Conclusion: The bio-psycho-social factors such as factors associated with the disease (e.g. anemia and bleeding), and the treatment (e.g. side effects of the drugs) and environmental and social factors are among underlying causes of some psychological problems in children with hemophilia.

Introduction

Hemophilia is an inherited bleeding disorder linked to the X chromosome resulting in a genetic deficiency in clotting factor VIII (hemophilia A) or factor IX (hemophilia B). It is classified as mild, moderate, or severe considering the percentage of the factor that is produced. Patients with hemophilia are at increased risk of joint bleeding (typically in the knees, elbows and ankles), intra peritoneal and intracranial hemorrhage, bleeding during and after surgery. There are also complications due to natural course of the chronic disease and the treatments the patients have to receive such as pain, anxiety, and depression. Children with hemophilia encompass a difficult life since they have to deal with chronic arthropathy followed by fatigue and limb limitations. One of the serious concerns of parents and health care systems is emotional and behavioral problems in children with hemophilia which gets more complicated with increasing age and may affect their quality of life.

Chronic diseases can lead to emotional, behavioral, and cognitive problems. Children with chronic diseases confront extreme stress, deprivation and serious limitations associated with their disease. Perceived stress can interfere with brain functions and may result in emotional and behavioral dysfunction. Hanson and colleagues indicated that difficult life circumstances in childhood can decrease hippocampus and amygdala volume which cause behavioral disorders. Children with hemophilia are often deprived of regular life activities because of the fear of recurring episodes of bleeding. The protracted stress can make them vulnerable to the brain atrophy which may result in behavioral problems. Prolonged bleeding causes a feeling of fatigue and weakness in children with hemophilia. The vitality of
children will be reduced and it will provide a context for developing a sense of anger and frustration. Children with hemophilia are susceptible to various infectious diseases and viruses such as hepatitis and AIDS. Parents of these children show overprotective behavior towards their children, resulting in anxiety and chronic depression in the children. Numerous studies have emphasized on low compliance of young patients with hemophilia to medical instructions. The emotional and behavioral problems disrupt the compliance with the treatment. Psychological consequences of hemophilia are worth studying. In this study, we aimed to assess the psychological status of children with hemophilia in three behavioral, emotional and cognitive aspects. Assessment of behavioral disorders and cognitive impairment in children with hemophilia by Wisconsin Card Sorting Test (WCST) is being investigated for the first time in our study comparing with healthy children.

Materials and Methods

The present research was a descriptive causal-comparative study. The study population consisted of all boys with hemophilia (aged 7-12 years) who referred to Children’s Hospital. Healthy children were selected as the control group from siblings of the patients. The convenience sampling method was applied for the study. The exclusion criteria were as follows: 1) existence of comorbidity, 2) history of psychiatric disorders, 3) having the ability to participate in psychological interventions, and 4) presence of any kind of intracranial hemorrhage. The effect of gender variable was also controlled due to differences in incidence of behavioral and emotional disorders between girls and boys. The sample consisted of 65 healthy children and 65 children with hemophilia who were assessed through Child Behavior Checklist (CBCL) and WCST. Informed consent was taken from the parents in advance explaining the motives of the study.

Child Behavior Check List (CBCL)

The Child Behavior Checklist (CBCL) is a report provided by the caregiver about children and is widely used in research and clinical practice. Generally, CBCL consists of the several following parts: 1) Academic performance assessment of children and adolescents in the fields of cognitive ability, training, and education issues, 2) Social skills assessment of children and adolescents for evaluating their adaptation with peers, siblings, parents and how they cope with challenges, and 3) Assessment of emotional and behavioral problems in children and adolescents.

The internal consistency of the CBCL was estimated using Cronbach’s alpha coefficients. Alpha coefficient of competence was relatively high and its range was between 65- 85% for CBCL. The test reliability based on Achenbach experimental assessment approach and by using Cronbach’s alpha was 0.89 for boys and 0.94 for girls and by using split-half reliability was 0.84 for boys and 0.87 for girls. The results of the construct validity supported the 8-factor structure of this scale by using factor analysis in Iran. In addition, convergent validity of this scale with “Junior Eysenck Personality Questionnaire (JEPQ)” and “Rutter behavioral problems questionnaire” was satisfactory.

If the index of discrimination power which is related to the variance and distribution scores reach higher than 90%, the scale will be appropriate. Obtained coefficients in the CBCL form were all at the high level. Based on the mentioned information about validity, it can be concluded that, CBCL is a valid tool in assessing behavioral and emotional problems and can be used with confidence by users.

Wisconsin Card Sorting Test (WCST)

The Wisconsin Card Sorting Test (WCST) which was first developed by Berg and Grant is a useful tool for studying cognitive deficits after brain injuries. WCST has been widely used as a neuropsychological test of “set-shifting”, i.e. the ability to display cognitive flexibility and abstract reasoning. Participants must maintain a concept found at the stage of testing in sequential conditions. When the classification rules change, they must change the previous concepts. Subjects were given a set of 64 cards which there were 1 to 4 symbols on them as triangles, stars and circles in 4 colors: red, green, yellow, and blue. Of course, there were no two identical cards. Four cards including a “red triangle”, two “green stars”, three “yellow Plus sign” and four “blue circles” were used as the main cards. The participant was told to match the cards based on 4 main patterns on the cards. After each response, participants received right or wrong feedback. In fact, whether a particular match is right or wrong. Scores obtained from this test included: number of incorrect responses, perseverative errors, and percentiles of achieved categories. Validity of this test has been reported 0.86 for measurement of cognitive deficits after brain injury.

The Kolmogorov-Smirnov test was applied to test normal distribution. It indicated normal distribution of Externalizing/Conduct, Inattention/Hyperactivity, Internalizing/Emotional and Social/Peer variables in experience-based scales. There was no normal distribution in other variables. Therefore, independent T-test was used to analyze the findings of mentioned variables and other variables were analyzed by The Mann-Whitney test.

Results

In this study, children with hemophilia (n=65) were compared to healthy children (n=65) in terms of cognitive, emotional, and behavioral problems (Table 1). The analysis of covariance demonstrated that there were no significant differences in age and socioeconomic status between the two groups. The minimum and maximum age in children with hemophilia was 7.5 to 11.2 years and in control group it was 7.8 to 11.6 years. Most of the children in both groups were in third and fourth grade of elementary school. Most parents were educated at the university level. The severity of hemophilia in most patients were of moderate type.

As shown in Table 2, there was no significant difference
between the two groups in average score of social behavior (P=0.430). The average score of academic performance in children with hemophilia was significantly lower than the healthy control group (P<0.01). Average scores of children with hemophilia in anxiety/depression, withdrawal/depression, somatic complaints, social problems, thought problems, attention problems, rule-breaking behavior, aggressive behavior, internalizing problems, emotional problems, anxiety, attention deficit hyperactivity disorder, and oppositional behavior were significantly higher than healthy children, while there was no significant difference between the two groups in physical problems (P=0.727).

According to Table 3, the result of t-test indicated that the average scores of activities and general competence variables in hemophilic children were significantly lower than control group and externalization and behavioral-emotional problems were significantly higher in children with hemophilia than healthy children (P>0.01).

As shown in Table 4, the average scores of the percentiles of achieved categories in children with hemophilia was significantly lower and preservation error variable was higher than healthy children.

According to Table 4, t-test results indicated that the average score of incorrect responses in children with hemophilia were significantly higher than healthy children (P>0.01).

**Discussion**

Based on the findings in our study, children with hemophilia indicated significantly lower scores than the control group in externalization/conduct, inattention/hyperactivity, internalization/emotions, and academic performance. This results were in line to the study of Coppola and colleagues who showed the academic achievement in children and adolescents with hemophilia were significantly lower than healthy children. Children with hemophilia would be more absent from the school due to occasional bleeding episodes. On the other hand, physical limitations prevent them from participating in school activities and do their homework same as their healthy peers. This could explain their poor academic achievement.
The results of our study showed that social behaviors in children with hemophilia do not differ from healthy children. Chiu and colleagues investigated the social functioning of children with hemophilia; they showed there were no significant differences in popularity and social acceptance of children with hemophilia compared to their healthy classmates.16 Chronic diseases cause increase in social sensitivity in the patients. They will also be concerned about judgment of their peers, so they comply with normal social behavior of their community to avoid negative self-image. As a result, they suppress their feelings and accept norms more easily.

Obviously, children with hemophilia compared with healthy children show more anxious/depressed and withdrawn/depressed conditions, somatic complaints, thought problems, attention problems and aggressive behavior, internalizing and externalizing problems and generally behavioral-emotional problems. Khair and colleagues conducted a qualitative study on 30 children with hemophilia. They revealed that hemophilia had a significant effect on family lives, educational issues, school and traveling plans. Most of them felt frustrated and expressed anger against the disease. Tryzapch and colleagues also assessed emotional functioning in hemophilic children.18 As their reports, children with hemophilia showed more problems in internalizing and anxiety/depression disorders. Children with hemophilia showed symptoms of depression more commonly compared to healthy children.19 There was also higher level of anxiety in hemophilic children which was significantly correlated with the parents’ attitude.20 Children with hemophilia in comparison to healthy group displayed more affection issues, attention deficit hyperactivity disorder and oppositional defiant disorder. A four-year longitudinal study indicated that children with hemophilia experienced more emotional problems; however, their quality of life improved over the time.21

Current research findings indicate a cognitive impairment in children with hemophilia through Wisconsin Card Sorting Test (WCST). WCST as a neuropsychological test displays frontal lobe functioning and cognitive flexibility (for assessment of executive functions, behavioral regulation, and social discourse). In a systematic review, Janual and colleagues demonstrated that hemophilia could not cause per se cognitive deficit and reduced IQ, but adverse effects of the disease could lead to serious cognitive and behavioral damages.22 So, we consider the bio-psycho-social factors as the main reason for behavioral and emotional problems.

Experience of pain and anemia effect on mood and temper of children with hemophilia (as bio factor). Over-parenting, lack of social learning and negative self-perception are involved in emotional problems (as a psychological factor). Children with hemophilia are less prone to socialization because they are afraid of participating in group activities. They are at increased risk for rejection by peers. So, social isolation may be a trigger for depression and anxiety (as a social factor). On the other hand, the child may feel anger followed by environmental and social deprivations. It would be expressed as aggressive behavior and conduct disorder.

In the study of Tryzapch and colleagues, children with hemophilia had not more externalizing problems compared to healthy children.23 However, in this study, externalizing problems in hemophilic children were more observed than healthy children. This difference may be explained by cultural and social issues, parenting styles and parents’ exaggerated understanding of child behavior problems.

The results of our study showed children with hemophilia demonstrate more impairment in executive functions. Although, previous studies have shown children with hemophilia who had a history of intracranial hemorrhage experienced impaired intelligence and visual perception, there was no cases of intracranial hemorrhage in our study. We showed cognitive function is influenced by the disease itself even in the absence of an intracranial hemorrhage.

Restriction in physical and social activities of hemophilic children from early childhood may interfere with development of cognitive processes. Also, many cognitive skills do not shape in these children properly.

### Table 3: T-test to compare activities, general competence, externalizing and general problems according to CBCL form

<table>
<thead>
<tr>
<th>Problem Scales</th>
<th>Children with Hemophilia</th>
<th>Healthy children (siblings)</th>
<th>t</th>
<th>P value</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>M</td>
<td>SD</td>
<td>M</td>
<td>SD</td>
</tr>
<tr>
<td>Externalizing/Conduct</td>
<td>16.24</td>
<td>5.13</td>
<td>6.28</td>
<td>2.18</td>
</tr>
<tr>
<td>Inattention/Hyperactivity</td>
<td>12.79</td>
<td>6.8</td>
<td>8.53</td>
<td>2.07</td>
</tr>
<tr>
<td>Internalizing/Emotional</td>
<td>17.6</td>
<td>4.09</td>
<td>11.62</td>
<td>2.72</td>
</tr>
<tr>
<td>Social/Peer</td>
<td>11.62</td>
<td>5.3</td>
<td>9.2</td>
<td>3.01</td>
</tr>
<tr>
<td>Total</td>
<td>49.73</td>
<td>13.18</td>
<td>20.93</td>
<td>7.22</td>
</tr>
</tbody>
</table>

### Table 4: T-test of Wisconsin Card Sorting Test (WCST) for participants

<table>
<thead>
<tr>
<th>Problem Scales</th>
<th>Children with Hemophilia</th>
<th>Healthy children (siblings)</th>
<th>t</th>
<th>P value</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>M</td>
<td>SD</td>
<td>M</td>
<td>SD</td>
</tr>
<tr>
<td>Categories Achieved</td>
<td>4.16</td>
<td>0.93</td>
<td>5.4</td>
<td>1.65</td>
</tr>
<tr>
<td>Failure to Maintain Set</td>
<td>3.67</td>
<td>2.5</td>
<td>5.17</td>
<td>4.06</td>
</tr>
<tr>
<td>Trial to first Category</td>
<td>2.36</td>
<td>2.91</td>
<td>5.32</td>
<td>3.45</td>
</tr>
<tr>
<td>Trial administered</td>
<td>15.8</td>
<td>4.31</td>
<td>19.2</td>
<td>2.43</td>
</tr>
<tr>
<td>Total trial Correct</td>
<td>16.1</td>
<td>3.55</td>
<td>20.43</td>
<td>2.64</td>
</tr>
</tbody>
</table>
Frequent absence from the school may be another factor that makes children to be less involved in school activities and it provides an inappropriate context for stimulation and training of intelligence and cognitive skills. This research suggests to perform future investigation of various psychological and neurological aspects in children with hemophilia in larger populations of the patients.

Conclusion

The bio-psycho-social factors such as factors associated with the disease (e.g. anemia and bleeding), and the treatment (e.g. side effects of the drugs) and environmental and social factors are among underlying causes of some psychological problems in children with hemophilia.

Conflict of Interest: None declared.

References


