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Social Work and Non-Pharmacological Treatment on Iranian Children with Hemophilia: A Systematic Review

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Abstract

Background: With the establishment of new technologies in the search for a cure for hemophilia, non-pharmacological treatments have had a more important role in increasing life expectancy, quality of life of children suffering from hemophilia disorders. The aim of the systematic review was a comprehensive understanding of Non-pharmacological treatment on Iranian children with hemophilia.

Materials and Methods: In this systematic review, articles published in national (Irandoc, Magiran, Medlib, and SID), and international databases (Medline [via PubMed], EMBASE, Web of Science, Scopus and Cochran Central Register of Controlled Trials) were searched without any time limitation till to June, 2019. The keywords of the search include (Child OR Children) AND (Hemophilia OR Hemophiliac) AND (Iranian or Iranian). To assess the quality of the articles intered into the search, we utilized the 5-item Jadad scale.

Results: Six studies were included in the systematic review. The achieved results suggested psychological training can alleviate the test anxiety, life expectancy. Yoga has a significant role in lessening the rate of bleedings, referrals to the haemophilia clinic, and the school non-attendance. Body vibration training has some benefits such as quadriceps strength, bone mineral density, and the functional capacity. Program management by nurse or own children have significant improvement on quality of life of children suffering from hemophilia disorders.

Conclusion: The achieved results suggested policymakers take heed of this novel treatment and non-pharmacological approaches such as psychological training, yoga, body vibration training and its effects on alleviating non-pharmacological treatment on children with hemophilia.

Key Words: Children, Hemophilia, Iran, Social Work, Treatment.

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1- INTRODUCTION

Hemophilia is a hereditary coagulation disease associated with the X chromosome with a prevalence of approximately 1 in every 10,000 people (1). The most common type of hemophilia is Factor 8 deficiency or hemophilia A, which accounts for approximately 80% of cases. Deficiency of factor 9 or hemophilia B accounts for approximately 20% of cases (2). Hemophilia is recognized as a blood coagulation inherited genetic disorder affecting nearly 18,000 males (1 per 7,500 men) in the United States. The chronic disorder, in which the deficient gene passed on through an X chromosome to the child, causes an impairment in the body's ability to make the blood clots during a lifetime (3). The disease, namely either haemophilia A or classic haemophilia, afflicts males almost exclusively, about 85% of which is due to the deficiency of factor VIII, the blood coagulating factor (4).

Early haemophilia symptoms including the prolonged bleeding following little scratches. Although it is generally assumed that such little but prolonged bleeding does not lead to the death, gradual progression of the disease causes more acute symptoms such as painful intra-articular bleeding, for instance, knee joint. Internal bleeding triggers the accumulation of blood inside the tissue and the joint, causes the joint to become inflamed and painful, and finally incapacitates it. Although the results of current gene therapy trials are very promising, there is no therapeutic treatment except the liver transplant for haemophilia A and B (5-8). Roughly speaking, the injection of factor VIII is the only efficient treatment in case of the severe and prolonged bleeding in the haemophilia patients. Since this factor is only obtained from the human blood plasma in a very low amount, it is exorbitant and inaccessible. Moreover, due to the frequent receiving of the blood products, the haemophilia patients are susceptible to the transfusion-transmissible viral infections: hepatitis B, hepatitis C, and HIV, to name but a few (4). The most life-threatening factors in the haemophilia patients are bleeding and chronic infections which cause several problems including prolonged physical disabilities, psychological stress and numerous financial issues in any period of one's life. Although the haemophilia patients make ultimate decisions on their healthcare management, they should also make sure that they do their best and are in the right position to express their social and economic problems including their disabilities, housing, and transportation issues, thereby, the authorities can address these issues in the process of policy design (9). Despite the wide range of complications in these patients, hemophilia patients can expect a near normal life and a healthy quality of life if treated appropriately (10-13). It should be noted that due to the high costs and lack of specialized care, access to alternative treatment on a large scale is limited to developed countries only (14).

In these countries, children today develop a relatively good musculoskeletal status, but these conditions are not yet possible for the majority of patients living in developing countries or with limited resources (14). In these conditions, hemophilia is managed by other available options that do not require expensive treatment products. In this regard, it should be borne in mind that when resources are limited, education as well as complementary therapies can be the cornerstone of hemophilia care (15). With the introduction of new technologies in the search for a cure for hemophilia, non-pharmacological treatments have had a more important role in increasing life expectancy, quality of life of children suffering from hemophilia disorders. The aim of the systematic review is a

comprehensive understanding of non-pharmacological treatment in Iranian children with hemophilia.

2- MATERIALS AND METHODS

Preferred Reporting Items for Systematic review and Meta-Analysis (PRISMA) checklist was used as a template for this review (http://www.prisma-statement.org/).

2-1. Data sources

The articles published in the national and international databases were enrolled in the systematic review and meta-analysis study. Two independent authors separately searched for the articles in the national (Irandoc, Magiran, Medlib, and SID), and international databases (Medline [via PubMed], EMBASE, Web of Science, Scopus and Cochran Central Register of Controlled Trials) without any time limitation till to June, 2019. Additionally, a manual search was conducted in Google motor engine, Google Scholar, and bibliography of correlated articles and reviews. The search query in Medline (via PubMed) is shown in **Table.1**. For further researches, a hand-search was carried out on the references list of the selected articles. Furthermore, some of the editorial boards of the national journals were approached in order to find grey literature.

Table-1: Search strategy for Medline (via PubMed).

((("child"[MeSH Terms] OR "child"[All Fields]) OR ("child"[MeSH Terms] OR "child"[All Fields] OR "children"[All Fields])) AND Hemophilia [All Fields] AND ("prescriptions"[MeSH Terms] OR "prescriptions"[All Fields] OR ("non"[All Fields] AND "drug"[All Fields] AND "prescription"[All Fields]) OR "Non drug prescription"[All Fields]) AND ("social work"[MeSH Terms] OR ("social"[All Fields] AND "work"[All Fields]) OR "social work"[All Fields]) AND (Iranian[All Fields] OR Iranian[All Fields]).

2-2. Included studies

Randomized controlled trials (RCT), clinical studies both randomized and nonrandomized either retrospective or prospective. Due to the limited number of published RCT in the literature other types of studies (semi-experimental, quasi experimental) were included. Pilot, preliminary and case report studies were not included due to limited sample size and higher risk of bias. Studies published in Persian and English till to June, 2019.

2-3. Study selection

Database search was done for possible studies, abstracts of the studies were screened for identification of eligible studies, full text articles were obtained and assessed and a final list of included studies was made. This process was done independently and in duplication by two reviewers and any disagreement was resolved by the 3rd reviewer.

2-4. Data extraction

Table.2 presents data extracted that contains several variables such as the first author of the article, study design, study population, intervention applied (type, duration of treatment and follow up), and main findings. Two reviewers collected the data independently collected data was combined and compared for accuracy any discrepancies were solved by a third reviewer.

2-5. Quality assessment

After searching and eliminating some reports and duplicates, two independent authors read the titles and abstracts, and then relevant studies were selected. Any disagreement was solved by discussion among the research team. To assess the quality of the articles entered into the search, we utilized the 5-item Jadad scale (16), consisted of randomization, randomization method, blindness, blindness method, and dropout/withdrawals and their motives (**Table.2**).

3- RESULTS

Six studies assessed the effect of non-pharmacological treatment on individuals with hemophilia (**Figure.1**). In the first study by Ghavidel Heydari et al., positive psychotherapy was run in eight sessions (two 120-minute sessions per week) followed by the subsequent set two months after ending the psychotherapy. Data analysis were carried out through the repeated measure analysis of variance. According to the results, positive psychotherapy helped considerably to alleviate test anxiety among the haemophilia patient students. Furthermore, the subsequent set corroborated the stability of the effects of this therapy (17).

In the second duplicate study conducted by the same authors, the results showed not only the significant effectiveness of positive psychotherapy on augmenting the life expectancy in haemophilia patients but also the consistency of this therapy in the subsequent set. This research underlined the prerequisite for training and the utilizing of positive psychotherapy as an innovative, risk-free and effective psychological therapy for the students suffering haemophilia (18).

In the third study by Beheshtipoor et al., yoga improved the mean scores of the quality of various life aspects, the rate of bleedings, the school non-attendance, and referrals to the haemophilia clinic ($p<0.001$). Hence, yoga with no trauma jeopardy approach may develop the haemophilia children's and juvenile's conception of the life quality. Additionally, this intervention apparently has a significant role in lessening the rate of bleedings, referrals to the haemophilia clinic, and the school non-attendance (19).

In the fourth study, 30 hemophilic children aged 9 to 13 years were picked and randomly designated to either study group that underwent the entire body vibration training i.e. 30-40 Hz, 2-4 mm of peak-to-peak vertical plate displacement for 15 minutes/day, 3 days/week for 12 weeks plus the conventional physical therapy program, or the control group in which merely a conventional physical therapy program was implemented. The subsequent results indicate some benefits such as quadriceps strength, bone mineral density, and the functional capacity. Children in the study group in comparison with those in the control group, made remarkable progress in all outcomes. Following the therapy, as seen in the results, the quadriceps peak torque was 70.26 and 56.46 Nm ($p<0.001$), lumbar spine bone mineral density was 0.85 and 0.72 g/cm2 ($p<0.001$), and the functional capacity was 325 and 290 m ($p=0.006$) for the study and control group, respectively (20). Fifth study evaluated the effect of case program management on quality of life of children in pre- school and found a significant difference between two groups on quality of life (21).

Sixth study assessed the effect of self-care behavior on quality of life of adolescence with hemophilia. Quality of life improved significantly in intervention group compared to control group (22).

Table-2: General characteristics of six studies (five studies and a duplicate study) included in systematic review.

Author, Year, Country, Reference	Duration of follow- up	Type of study	Intervention	Control	Treatment	Control	Main findings
Beheshtipoor et al., 2015, Iran, (19)	14 weeks	Semi-experimental with pre- and post-test	Youga.	-	27		After the intervention, significant differences were observed in the mean scores of quality of life dimensions and the number of bleedings, school absences, and referrals to the haemophilia clinic (P<0.001).
Ghavidel Heydari et al., 2018, Iran, (17)	8 sessions (two 120 minute sessions weekly)	Quasi-experimental	Positive psychotherapy workshop was held.	No treatment	30	30	Positive psychotherapy on increasing Life expectancy and anxiety.
Agha Seyed Mirza et al., 2013, Iran, (21)	Three months	Semi experimental	Case management program.	No treatment	26	26	Quality of life improved significantly in intervention control compared to control group.
Asgarpour et al., 2007, Iran, (22)	Two educational sessions of self -care (45 minutes)	Quasi experimental	Two educational sessions of self-care.	No treatment	27	29	Difference was significant between two groups in after intervention (P=0.001).
El-Shamy, 2017, Iran, (20)	Body vibration training for 15 minutes/day	Randomized clinical trial	Body vibration training plus the conventional physical therapy program.	Conventional physical therapy program	15	15	Difference was significant between two groups in after intervention.
Ghavidel Heydari et al., 2019, Iran, (18)	8 sessions (two 120 minutes sessions weekly)	Quasi-experimental	Positive psychotherapy workshop was held.	No treatment	30	30	Positive psychotherapy on increasing life expectancy in patients with hemophilia.

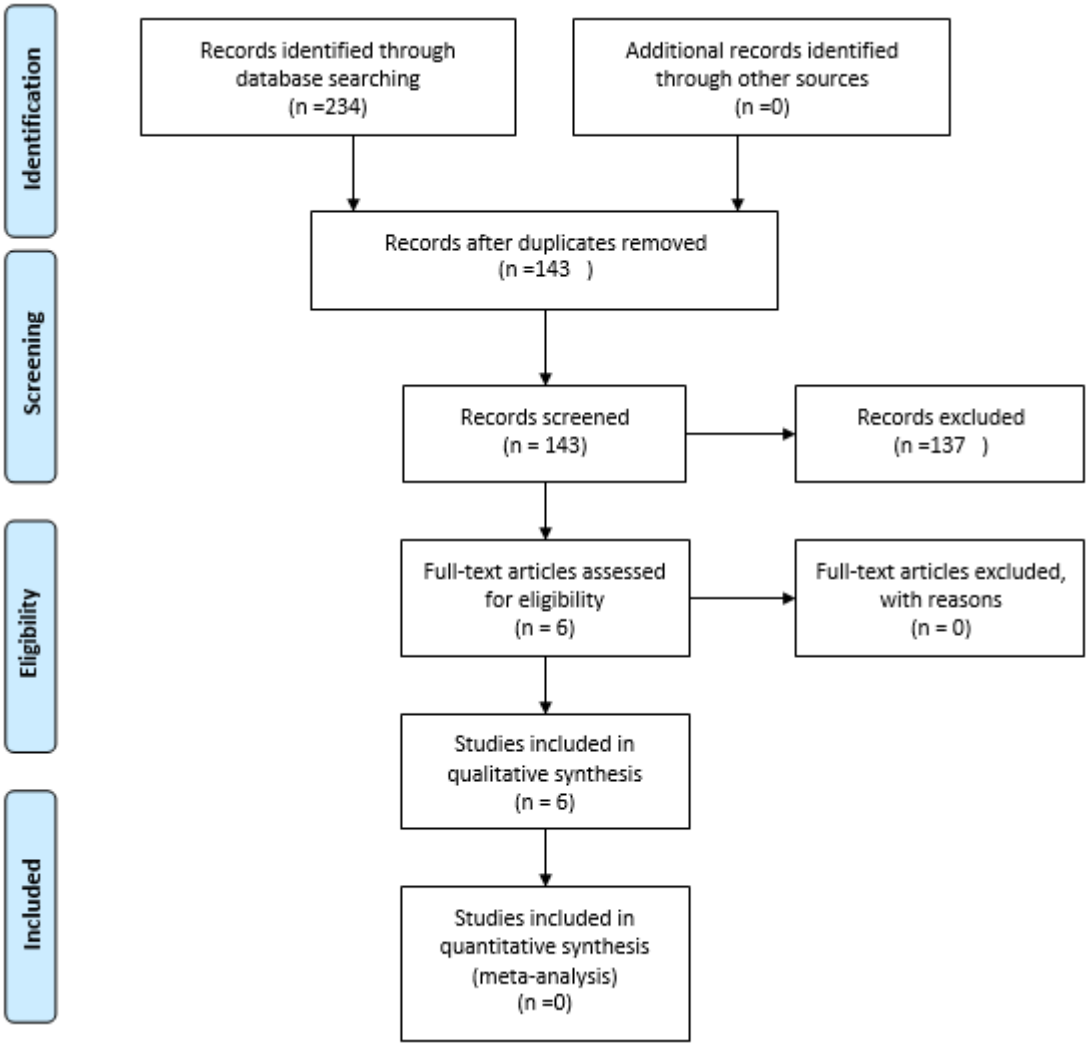

Fig.1: PRISMA flowchart.

4- DISCUSSION

The aim of the systematic review was a comprehensive understanding of non-pharmacological treatment in children with hemophilia. The introduction of new technologies in the treatment of hemophilia underlines the role of social workers in the healthcare and non-pharmacological treatment in prolonging the life expectancy process in patients with bleeding disorder. Among the eastern Mediterranean region, Iran is one of the major centers for the prevalence of β-Thalassemia. Regarding the high consanguinity among population, it is estimated that there are between two and three million β-Thalassemia carriers and 25,000 patients in Iran (24, 25). Thalassemia carriers are today present worldwide with high frequencies mainly in the endemic countries of Africa, the Middle East, the Indian subcontinent, Southeast Asia, and the Mediterranean region (26-28). Thalassemia is a common disease in Iran with higher frequencies in the north and south parts of the country (29-31). The achieved results suggested policymakers take heed of this novel positive psychological training and its effects on alleviating the test anxiety (17), and the life expectancy (18). Yoga has a significant role in lessening the rate of bleedings, referrals to the haemophilia

clinic, and the school non-attendance (19). The subsequent results indicate body vibration training has some benefits such as quadriceps strength, bone mineral density, and the functional capacity (20). Program management by nurse or children showed significant improvement in interventions group compared to control group level on quality of life (21, 22).

4-1. The Social work and the prenatal genetic counseling in the chronic haemophilia

Although the physicians and the geneticists traditionally provide the genetic counseling for the chronic diseases and the risks of deficient gene transfer, the social workers and other trained people play an important role in trying to deal with psychological and social issues followed by the non-fertility recommendation in couples who have the gene-related problems or abort the child with disorder. Since the social workers are familiar with the family' emotional, financial, and the social resources, they can provide these opportunities through the emotions, the attitudes and the human values as well as family therapy in their communication with the clients (9, 23).

4-2. The social work with haemophilic children

Children spend more time on learning, recreation, playing and communicating with their peers. But children with the blood disorder face a variety of stress factors, including the suffering from their physical conditions and the subsequent harassment of peers. Hence, the social worker skills in working with these children should be aimed at empowering the children and their families with the following objectives:

- The preliminary long-term prophylaxis before the age of 1 or 2 years and before joint damage at the school and ensuring the safety checking in the family.

- A new generation of parents of the children with haemophilia are required to have factor VIII injections at the treatment centers, 2 or 3 times a week; conversely the older generation of parents had a tendency toward using the blood products. Therefore, the social workers in this field played a role in chaing the attitude of of people so they prefer the pain tolerance over the use of the blood products.

- Addressing the emotional state of the clients.

- Family education, the teaching of various types of parenting styles for these people.

- The financial advice and adequate support from the family.

- A serious challenge for many haemophilic children leading to the negative consequences of social isolation is caused by the physical differences, motion constraints and the peers' attitude about these children. Spotlighting the individual talents and brain functions of the child, such as playing chess and improving the child's verbal abilities and social skills, could help to avert the issues, which haemophilic children are faced with.

- Most of the children and their families tend to conceal their child's disease. The social workers are suggested to help the parents apprise the school staff to diagnose children's disease through the differences and constraints these children deal with in their developmental stages.

- Focus on teaching the useful skills in the confrontation with the peers.

- Seek an opportunity for the peers' connection: harassment of the older children could be overcome by one close friend of the patient. Since the parents play a significant role in the child's social adaptation, the social workers should pay heed to the fact that parents' efforts to create the strong social networks and emphasize on the need for these interactions such as the physical care of the child is precious (3, 9).

5- CONCLUSION

The achieved results suggest policymakers take heed of this novel treatment and non-pharmacological treatment such as psychological training, yoga, body vibration training and its effects on alleviating the test anxiety, the life expectancy, lessening the rate of bleedings, referrals to the haemophilia clinic, and the school non-attendance and some benefits such as quadriceps strength, bone mineral density, and the functional capacity.

6- CONFLICT OF INTEREST: None.

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