

## ABSTRACT

Hemophilia is an incurable hereditary bleeding disorder due to the absence or deficiency of clotting factors in the blood. This disease was traditionally classified as mild, moderate, and severe based on the level of coagulation factors. Oftentimes, Hemophilia causes flexion deformities of the elbows, knees, and hips interfering with the individuals' daily activities. The aim of this review was to examine the literature which addressed quality of life for adolescents with Hemophilia. A literature search was conducted using CINAHL and EBSCO databases using the keywords "blood disorders and adolescents". Search refinement with keywords "quality of life and hemophilia and adolescents and nursing" resulted in 30 articles. The search was again refined to include only full text articles dated from 2014 – 2019. The result was six articles. However, one article was eliminated. Review of the literature revealed Hemophilia has an impact on the Health related quality of life (HRQoL) of patients.

## INTRODUCTION

Bleeding disorders are a group of disorders that have the inability to form blood clots properly. Improper clotting can be caused by defects in blood components such as platelets and/or clotting proteins, also called clotting factors. Hemophilia is a rare group of bleeding disorders resulting from congenital inherited deficiency, dysfunction, or absence of specific coagulation proteins or factors that cause longer healing, may damage organs, muscle tissue, and joints. Hemophilia is transmitted as an x-linked recessive disorder only about sixty percent of affected children have a positive family history for the disease. Though one-third of hemophilia cases may be caused by a gene mutation. There are several types of bleeding disorders, two common forms are Hemophilia A (classic) Factor VIII and Hemophilia B (Christmas disease) Factor IX. This rare bleeding factor prevents the blood from correct clotting which leads to prolonged bleeding after injury, surgery or physical trauma and can become deadly if not treated correctly. Bleeding disorders lessen the physical form of health-related quality of life (Beheshtipoor et al.).



**Hemophilia**

## METHODS

A literature search was conducted using CINAHL and EBSCO host databases using the keywords "blood disorders and adolescents". The search yielded 1600 articles. Search refinement with keywords "quality of life and hemophilia and adolescents and nursing" resulted in 30 articles. Refinement of the search occurred using keywords "hemophilia and adolescents and quality of life" and include only full text articles that were dated 2014 – 2019. The result was six articles. However, one article was specific for children and eliminated from the review which resulted in five articles.

## RESULTS

### Article 1

Owaidah et al. (2018) conducted an epidemiological based survey among adolescents and young adults (1901 [49%] boys, 1980 [51%] girls) in Riyadh city regarding bleeding symptoms. Of these, 1849 (47.6% responded "Yes/Positive" for at least one question about the bleeding symptoms. The Most common bleeding symptom was epistaxis (19.7% of the sample population) detected in Phase I of the study. A tandem survey was conducted among 525 adolescents, who responded "Yes/Positive" to any one of the questions in to any one of the questions inquiring about bleeding symptoms. In this study, they reported for the first time the prevalence of bleeding symptoms in a representative sample of Saudi adolescents and young adults (Owaidah et al.).

### Article 2

Neuner, et al.(2016) conducted a cross-sectional study comparing the Health-related quality of life (HrQoL) amongst children and adolescents with a Hereditary Bleeding Disorder to that of children and adolescents with stroke/TIA. Participants were admitted into 6 specialized treatment centers in Germany. The population was initially 354 participants, 37 of which were older than 17 years old. (10.5%). Of the remaining 299 study participants 23 did not fill out the revised KINDer Lebensqualitätsfragebogen (KINDL-R) questionnaire. The final data set consisted of 144 patients 74 who were patients with HBD(51.4%) and 70 who were patients who stroke or TIA(48.6%). In conclusion Group 1 showed no differences in self reported HrQoL between patients and siblings and between patients and peers. While Group 2 showed differences between patients with stroke/TIA and their healthy peers regarding self-worth and friend-related well being (Neuner et al.).

### Article 3

McLaughlin et al. (2017) conducted a cross-sectional study among adolescents and young adults with bleeding disorders. The purpose of their study was to investigate the relationships between pain management and quality of life while increasing awareness, knowledge of treatment and living with rare bleeding disorders. An online 36-item Short Form Health Survey (SF-36) approved by Munson Medical Center institutional review board, and IMPACT quality

## RESULTS cont.

of life survey was de-identified prior to data collection and analysis of while associated factors. Study participants were participants 13 to 25 years of age, able to read, write, and speak English; and had a diagnosis of hemophilia A, hemophilia B, or von Willebrand disease. Information was delivered at major US hemophilia meetings, treatment centers and a designated Facebook page from April to December of 2012. The final regression model shows physical component score (PCS) and the median mental component score (MCS) amongst 108 adolescents and young adults with hemophilia A, B and von Willebrand disease. Half of participants were males with hemophilia and von Willebrand disease. However the opposite was true for females with von Willebrand disease than hemophilia (McLaughlin et al.).

### Article 4

Beheshtipoor et al. (2015) conducted a semi-experimental study at the children's hospital in Shiraz, Iran. It studied the effects of yoga on the quality of life in children and adolescents with Hemophilia. The study was performed on 27 boys with Hemophilia that were divided into two groups. The quality of life was assessed using the Haemophilia Quality of Life (Haemo-QoL) questionnaire. Yoga intervention was performed for 14 weeks and the data was entered in the Statistical Package for Social Science (SPSS) statistical software version 18 and analyzed using non-parametric Friedman test. The inclusion criteria of the study included the ability to read and speak Persian having no prior experience in yoga. None of the 27 boys dropped out of the yoga study. The results of the Friedman test indicated these boys showed a significant difference in the quality of life for a reduction in the mean number of bleeding events ( $\chi^2=44.07, P<0.001$ ), referrals to the Hemophilia clinic ( $\chi^2=18.28, P<0.001$ ), and school absences ( $\chi^2=40.13, P<0.001$ ) throughout the assessment stages. The study concluded support for the safety and effectiveness of the adolescents performing yoga, which resulted in the increase of their quality of life (Beheshtipoor et al.).

### Article 5

Zhang et al. (2019) conducted a prospective cohort longitudinal study that was started in August 2014 in Nanjing, China, Children's Hospital affiliated with Nanjing Medical University, and the follow-up was completed in January 2018. The study was among 42 children with Hemophilia and their parents. Among these 42 subjects, 37 had Hemophilia A and the other 5 had Hemophilia B. Children aged 7 years old and older, if literate, were asked to complete self-report versions of the Canadian Haemophilia Outcomes–Kids' Life Assessment Tool (CHO-KLAT). Also, their parents were asked to complete a proxy CHO-KLAT on their behalf. The Chinese version has an additional unique Socio-Economic Context (SEC) module, which consists of 9 questions covering information about stigma, opportunities for stable employment and access to treatment due to cost, etc. SEC scores have the same range as those of the original CHO-KLAT (Zhang et al).

## DISCUSSION

Findings from this review indicate children and adolescents with Hemophilia have increased financial burden due to expensive treatment. There was also a physical activity restriction in order not to suffer any injuries, but this also depends on the severity of the disease and the rate of bleeding during bleeding episodes. The findings also revealed engaging in yoga had a positive impact on adolescents with Hemophilia quality of life (Beheshtipoor et al., 2015) Pain management and education also proved effective in managing Hemophilia in adolescents (McLaughlin, et al.). Studies show that there also is an increased prevalence of Hemophilia in male children and adolescents than females and also that there is a decrease in physical and mental activity in males and no change in female children and adolescents with the disease (McLaughlin et al., 2017)

### Future Research Questions Generated from the Review

1. What is the difference in quality of life measures for adolescents with or without Hemophilia?
2. What diversionary activities improve the health related quality of life in adolescents with Hemophilia residing in the United States?
3. What factors predict improved quality of life for US adolescents living with Hemophilia?

## CONCLUSION

Adolescents with Hemophilia have a decreased quality of life due to physical regression and restrictions which impacts their physical and mental well being. Yoga, pain management and education improved quality of life for adolescents (McLaughlin, et al.). Understanding the impact of Hemophilia on adolescents is essential to providing nursing care that will improve health outcomes and adolescents overall quality of life. Studies were limited to primarily international communities.

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