INNOVATIONS

Evaluation of quality of life of children with haemophilia and their parents coping at haemophilic society: a systematic review

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Abstract: Background: A child's diagnosis of hemophilia is actually life-changing in its consequence on both the child and the parents, who face a future of controlling bleeding risk during which trying to supply the child and his siblings with as normal a life as possible. **Aim:** The aim of this systematic review was to summarize quality of life of children with hemophilia and coping approaches used by parents of hemophilic children, identify which tools are most frequently used to measure quality and coping strategies in parents of hemophilic children and report on outcomes of quality of life of hemophilic children and coping strategies in parents of hemophilic children. **Method:** we searched for articles indexed in PubMed, Web of science, Psyc INFO, and CINAHL database using a combination of expressions including "Quality of life" AND "Hemophilic children" OR "Coping AND ''Parents". **Results:** Six empirical studies were identified as relevant to our research. Several types of tools are used to assess quality of life of hemophilic children and coping strategies in parents of hemophilic children. We found that the relation between quality of life and hemophilic childrenparents of hemophilic children used more functional coping styles. The most widely used strategy parents of hemophilic children appears to be positive coping behaviors. **Conclusions:** This review underlines that quality of life is impaired in hemophilic children and in hemophilic parents which many aspects of life are affected.However, providing care can be rewarding and program of support, education and suitable treatment improve the well-being of quality of life of children with hemophilia and their parents coping at hemophilic children.

Keywords: 1. Quality of life **2**, hemophilic children **3**. coping and Parents

1. Introduction:

A child's diagnosis of hemophilia is actually life-changing in its consequence on both the child and the parents, who face a future of controlling bleeding risk during which trying to supply the child and his siblings with as normal a life as possible. **(Kate Khair and Steve Chaplin, 2016)**Hemophilia is a hereditary hemorrhagic disorder identified by deficiency or dysfunction of definite coagulation protein factors. Recurrent joint and muscle bleeds show to severe and growing musculoskeletal impairment. **(KaterinaRatajova et al., 2019)**Hemophilia is an x-linked recessive genetic diseaseidentified by low levels of important clotting factors. The two common types of hemophilia are factor VIII deficiency and factor IX deficiency. Worldwide, hemophilia affects 400,000 people, with an approximate prevalence of 1 in 5000 male live births for hemophilia A and 1 in 30,000 live births for hemophilia B. **(RohanPratap et al., 2020)**The feature of hemophilia are intramuscular and intra-articular bleeds. Some children evolvetarget jointsidentified by multiple bleeds into the same joint. This can appear to the demolition of the joint surfaces or haemophilicarthropathy.**(C. R. Broderick et al., 2010)**The most clinical results in haemophilia are for haemophilia B, where intravenous infusion of an adeno-associated viral vector encoding factor IX (FIX) under the control of a liver-restricted promoter has reported in expression of FIX at plateau levels ranging from 1% to 6%, for periods of 2 years, in 6 adult males with severe haemophilia B.**(Paola Giordano et al., 2013)**Hemophilia is the most usualof the severe bleeding disease and enjoys the most effective and safe treatment among the prevalent monogenic disorders; although, if not correctlycontrol since early infancy, it can appear to chronic disease and lifelong disabilities.**(Marta Bertamino et al., 2017)**

1.1 Review aims

This review explores to provide a summary of quality of life of children with hemophilia and coping approaches used by parents of hemophilicchildren. Thus, the main aim of this systematic review was to (1) summarize quality of life of children with hemophilia and coping approaches used by parents of hemophilic children (2) identify which tools are most frequently used to measure quality and coping strategies in parents of hemophilic children and (3) report on outcomes of quality of life of hemophilic children and coping strategies in parents of hemophilic children.

2. Material and method

2.1 Study selection and data collection processes

A comprehensive search of electronic databases including PubMed, Web of science,Psyc INFO, and CINAHL database was conducted as part of a systematic review examining outcomes of quality of life of hemophilic children and coping strategies in parents of hemophilic children. All

databases searches were using a combination of the following free-text terms: “Quality of life” AND “Hemophilic children” OR “Coping AND ‘‘Parents”.In the initial stage, duplicates were excluded and reference lists of relevant articles were examined to identify additional studies meeting the criteria for inclusion. After this first literature search, titles and abstracts were screened by reviewers to identify those meeting inclusion criteria.

2.2Eligibility criteria

The studies included in this review met the following criteria: (1) Quality of life of hemophilic children and coping strategies in parents of hemophilic children, (2) An intervention study focused on quality of life of hemophilic children and coping strategies in parents of hemophilic children, (3) Used quantitative research includes a non-intervention or pre-post comparison group to examine quality of lifeof hemophilic childrenand coping strategiesin parents of hemophilic childrenand (4) The paper were published in a peer – reviewed journals with original research articles.Paper were rejected if they:(1) Without mother coping strategies, (2) were qualitative studies, review papers, case reports /case series, thesis/dissertations and (3) were not published in English.

2.3Data extraction

A data extraction tool was used to systematically record data from included studies: (1) Study characteristics, author type of study design, year and country where data were collected, (2) Characteristics of children:number of children, mean age of children and diagnosis, (3) Characteristics of parents: number of parents, mean age of parents, (4) Tools used to measure coping enhancement and (4) Findings

2.4Study quality assessment

Study quality was evaluated using the quality assessment tools from: (1) The STROBE reporting guidelines for observational studies and (2) The critical review form for quantitative studies. Each question could be answered complete (score= 2), partially (score=1) and imprecise (score=0). A total score was calculated for each study. Studies were then rated as poor (total score less than 12 points), fair (total score between 13 and 24 points), good (total score between 25 and 30 points), or excellent (total score between 30 and 36 points), based on the scores obtained. Studies were rated independently by third reviewers.

3. Results

Our search strategy yielded 339 studies (PubMed n = 137, Web of Science n = 101, PsycINFO n= 76 and CINAHL n = 25). After excluding duplicate publications, we identified 208 potential

articles. In the screening phase, titles and abstracts of all identified studies were examined. This led to the exclusion of 125 studies, as they were not deemed suitable for the present review. Consequently, 49 studies were selected for the eligibility phase. Out of these, 16 studies were excluded because they did not meet selection criteria. Finally, 6 empirical studies were ultimately identified as relevant to our research **(Table 1).**

3.1. Study quality

The quality rating was "fair" for two studies**(JiePeng et al, 2017; Anna Marie D Espaldon and Flerida G Hernandez 2014)**and "good" for the remaining four **(GustiAyuTrisnaWindiani et al, 2020; Linda MyrinWestesson et al, 2019; Sayed Hamid Mousavi et al, 2019; V. Manikandasamy et al, 2017)**

3.2. Study characteristics

The main methodological features and general characteristics of all reviewed studies are summarized in **Table 1.** All studies are cross-sectional studies **(GustiAyuTrisnaWindiani et al, (2020); Linda MyrinWestesson et al, (2019); Sayed Hamid Mousavi et al, (2019); V. Manikandasamy et al, (2017); JiePeng et al, (2017); Anna Marie D Espaldon and Flerida G Hernandez (2014)).** With respect to research methodology, there were no randomized controlled trials and interventional study. All studies were published from 2014 to2020. Three of the included studies had been conducted in Indonesia, Sweden, Iran, North India, China and Philippines.

Table1: Participant Characteristics

Author(year)	Type of study design (Nation)	Number of children (n)	Age of children (years)	Number of parents (n)	Quality of life and Coping measures	Findings
GustiAyuTrisnaWindianiet al, (2020)	Cross sectional study (Indonesia)	22	4-18	22	PedsQL Generic Core Scales version 4.0 inventory	Total PedsQL score in hemophilic children showed a significant difference inchild and parent-proxy reportat $p<0.05$.
Linda MyrinWestessonet al, (2019)	Cross sectional study (Sweden)	102	0-17	102	Hemophilia associated caregiver Burden scale	Higher burden was seen in parents of children atp = 0.010
Sayed Hamid Mousaviet al, (2019)	Cross-sectional study (Iran)	65	8-16	65	Persian version of Haemo-QoL Questionnaire	The mean Quality of life score was 75.9 ± 17.4. Mostly (80%)the children were suffered from hemophilia A.
V. Manikandasamyet al, (2017)	Cross sectional study (North India)	51	4-12	51	Hemo-QoL questionnaire	Perceived impact on family, poor physical health, sports and school had (77.3±14.7), (62.9±29.8), and (53.8±22.8) the highest negative impact on Quality of life.
JiePeng et al, (2017)	Cross-sectional Interview study(china)	158	3-18	158	Coping Health Inventory for Parents	There was a significant difference in coping behaviors in maintaining a normal familylife by parents' education and income at p <0 .001.
Anna Marie D Espaldon and Flerida G Hernandez(2014)	Cross sectional study (Philippines)	51	4-16	51	Haemo-QoLandTransformed Scale	The total mean Transformed Scale Score was 28.39 ± 4.76 which reflecting the good Quality of life.

3.3. Characteristics of children and mothers

These 6 studies, 499 werehemophilic children with parents. The ages of hemophilic children were between 0 to 18 years.

3.4 Quality of life and Coping measures of hemophilic children and parents

This review of Quality of life and Coping measures showed**GustiAyuTrisnaWindiani et al, (2020)**used the PedsQL Generic Core Scales version 4.0 inventory; **Linda MyrinWestesson et al, (2019)**Hemophilia associated caregiver Burden scale; **Sayed Hamid Mousavi et al, (2019)**investigated Persian version of Haemo-QoL Questionnaire; **V. Manikandasamy et al, (2017)**used Hemo-QoL questionnaire; **JiePeng et al, (2017)**administered Coping Health Inventory for Parents; **Anna Marie D Espaldon and Flerida G Hernandez (2014)**adopted Haemo-QoL and Transformed Scale.

3.5 Outcomes of quality of life of hemophilic children and coping strategies in parents of hemophilic children

3.5.1 Relationship between quality of life and hemophilic children

Three studies analyzed the relation between quality of life and hemophilic children.**GustiAyuTrisnaWindiani et al, (2020)**adopted physical (71.8%), emotional (81.4%), social (85.0 %), and school function (73.1 %). In cross-sectional study, **Sayed Hamid Mousavi et al, (2019)**showed that there was a significant correlation between age and quality of lifescores at $r = 0.8$ and $P = 0.02$. **Anna Marie D Espaldon and Flerida G Hernandez (2014)**reported that most impairment in the subscale of Family(43.75 ± 36.8), in Sports and School subscale (58.2 ± 18.77 and 59.27 ± 17.46), in Attitude (6.25 ± 7.60) and in Treatment subscale (12.5 ± 15.26 and 23.99 ± 11.02).

3.5.2 Relationship between coping styles and parents of hemophilic children

Among selected studies, two studies reported the relation between coping styles and parents of hemophilic children. **Linda MyrinWestesson et al, (2019)**examinedhigher burden was seen in parents of hemophilic children with past or present inhibitors,in parents of younger hemophilic children, if a member of family managedthe clottingfactor and in parents of hemophilic children with overweight or obesity.**JiePeng et al, (2017)**found that parents of hemophilic children involved in positive coping behaviors of Children's illness and physical conditions, Children's treatments to control andEconomic problems these stressors. There was a significant difference between the coping behaviors and education and income at $p < .01$.

3.5.3 Relationship between coping and quality of life

V. Manikandasamy et al, (2017)reported thegreatest negative outcome on quality of life of Perceived impact on family (77.3±14.7), poor physical health (62.9±29.8), sports and school (53.8±22.8).Support from friends, family, and other persons appeared to have provided positively as regards the quality of life. Parents of older hemophilic children (40.8±14.2)had greater mean subscale scores as compared to parents of younger hemophilic children (23.7±33.0) at p value 0.018. Under Sports (48.2±20.7)and school (34.1±13.8) subscale was perceived at P value 0.045.

4. Discussion

This review was mainly aimed at exploring the Evaluation of quality of life of children with hemophilia and their parents coping at hemophilic society. The quality appraisal of the reviewed papers showed that, theycontentedmost of the required criteria includerelevanceof the topic; methodological qualityand analysis of the results and accordingly impact were agreeable.The reviewed articles which includedquality of life and hemophilic children, coping styles and parents of hemophilic childrenshowed the outcome of raising hemophilic children on the use of quality of life and coping measures.The most commonly used quality of life by hemophilic children seems to be physical, emotional, social, and school function **(GustiAyuTrisnaWindiani et al, (2020); Gustavo CambraiaTrindadeet al, (2019);Sylvia von Mackensenet al, (2018); John M. McLaughlin et al, 2017;Jiat-Ling Poon et al, 2014; Adriana Aparecida Ferreira et al, 2013;SinghM et al, (2020)).**Similarly,**Hee Jo Baeket al, (2019); Alexandra FuenmayorCastan et al, (2017); ShahparBagheri et al, (2013) andA. Dsouzaet al, (2020)** suggests that Sports and leisure, family planning, perceived support, friends and dealing are the higher impairment of Health-related quality of life of hemophilic children. In contrast with previous studies showed that quality of life was significantly affected in children with bleeding episodes and presence of target joints**Mohammad Salmaan et al, (2016); Heng Zhang et al, (2019).**In addition, two reviewed studies showed thatcoping styles and parents of hemophilic children**(Linda MyrinWestesson et al, (2019); JiePeng et al, (2017)).** These findings suggest thathigher burden was seen in parents of hemophilic children and Parents involved in positive coping behaviors of Children's illness and physical conditions, Children's treatments to control andEconomic problems.Previous authors have reported that Mothers hemophilic children depression and anxiety scores are related to hemophilic children depression, anxiety **MerveCikili-Uytun et al, (2020); Tyler W Buckner et al, (2019);FatemehMakki and Zahra Nikmanesh (2018).**Similarly, **Kate Khair and LemuelPelentsov (2019);Maria Elisa Mancuso et al., (2020)** described Parentsof hemophilic children showed a good understanding,

experiencing significant emotional issues and financial concerns of their child's condition.**V. Manikandasamy et al, (2017)** reported that coping and quality of life of hemophilic children. Previous studies reported that improvement in knowledge scores immediately after intervention**S. Phadnis and A. Kar (2016).**

Finally, our review highlights the paucity of published studies on quality of life of children with hemophilia and their parents coping at hemophilic children. This would be a fruitful area for further research, in order to establish whether parents coping strategies varyaccording to hemophilic children. There is also a certain lack of data on follow-up measurements and transfer and generalization outcomes of parents training on coping strategies. We especially excluded qualitative studies, even though they do provide highly to the literature in this field, because their findings cannot be expanded to large people with the similar degree of reliability as quantitative analyses.

5. Conclusion

Our review shows that quality of life of children with hemophilia and their parents coping at hemophilic children. Overall, this evidence showed that quality of life is impaired in hemophilic children and in hemophilic parentswhich manyaspects of life are affected. However, providing care can be rewarding and program of support, education and suitable treatment improve the well-beingof quality of life of children with hemophilia and their parents coping at hemophilic children.

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