GUIDELINE PROGRAM INTERVENTION FOR MOTHERS REGARDING TO CARE OF THEIR CHILDREN SUFFERING FROM OSTEOGENESIS IMPERFECTA

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ABSTRACT

BACKGROUND: Osteogenesis Imperfecta (OI) is a chronic genetic condition affecting children and described as “Brittle Bone disease.” This condition is expressed by low bone density and characterized by frequent fractures with or without trauma, pain, altered growth, deformities and disability.

AIM: to evaluate the effect of guideline program intervention on mothers regarding to care of their children suffering from osteogenesis imperfecta (OI).

METHODS: A purposive sample of 80 mothers and their children suffering from Osteogenesis Imperfecta (OI) who attended the genetics department, Children Hospital, Ain Shams University, Cairo, Egypt from January 2019 till the end of February 2020. Data collection included an interviewing questionnaire; mothers reported practices and quality of life scale for both mothers and their affected children.

RESULTS: Maternal knowledge showed significant increase to 78.6% after the intervention program compared to only 42.5% pre-interventional program. Their daily practice also improved significantly to 82.9% compared to 48.8% pre-intervention. Interestingly, quality of life high scores has increased to 74.3% compared to only 22.5% pre-intervention. Additionally, there was positive correlation between all variables of the study (mothers’ knowledge, reported practices, quality of life scale for both mothers and children).

CONCLUSION: Intervventional program is effective in improving caregiver’s knowledge and practice about OI and subsequently improving the scores of quality of life for both children and their caregivers. Enforcement of this program is highly recommended to all families with OI.
I. INTRODUCTION

Osteogenesis Imperfecta (OI) known as brittle bone disease, is a genetic disorder of connective tissue which is characterized by bones that fracture easily from little or no apparent trauma. However, the multiple fractures will lead to progressive bone deformities, short stature and restricted mobility [1].

OI affects an estimated one individual per 10–20,000 worldwide, making it one of the rarest skeletal dysplasia [2]. Types one and four are the most common forms of OI, affecting 4 to 5 per 100,000 people. In autosomal dominant types affected parents have a 50% chance to pass the disease to their children [3].

Children with OI go through the same ageing process and subjected to the same common disease as the general population but they have unrestricted or highly compromised mobility and quality of life. Therefore, the need for increased awareness of potential bone fragility is highly indicated [4].

Nurses play a key role in guidance and support of these children and their families. They coordinate the involvement of the multidisciplinary team helping to instigate a policy of continuity of caring of children and their caregivers. This is why the earliest nursing intervention should be at the time of diagnosis [5].

Nurses should be able to provide the family with general information about the child condition and encourage the parents to participate in child’s care. They should encourage confidence and increase understanding of handling and management. Also, it is important to help families find support for coping with their feelings and referral to family social service [6].

AIM OF THE STUDY

The present study aimed to evaluate the effect of guideline program intervention on mothers regarding to care of their children suffering from OI.

II. METHODS

In the present study, quantitative and evaluative approaches were used, research design was quasi experimental: One group pre-
test - post test. Study was conducted at Genetics department, Children Hospital, Ain Shams University, Cairo, Egypt.

The sample size included 80 mothers and their affected children in pre- test, but it decreased to 70 in post- test. The quality of life scale of children applied on 44 child in pre -test, their age ranged from 5 to 15 years, but decreased to only 40 post- test.

**Tools of data collection: pre /post intervention:**

Data was collected through using the following tools pre /post intervention:

1. **An interviewing questionnaire sheet:** it consisted of three parts, the first part was related to characteristics of the study subjects (mothers and their children), second part was related to the past and present medical history of children suffering from OI. While the third part was concerned with the mother’s knowledge regarding OI such as meaning, causes, clinical manifestations, types of conventional treatment, medication used for treatment of OI and their precautions and side effects. Also, mothers' knowledge about the importance of compliance of medical treatment and preventive measures for the expected complications for OI.

   **1. Scoring system:**

   Mothers' answers were checked with the model key answer and scored one degree for the "correct answer" and zero for "incorrect answer".

   Scores of questionnaire were summed up and accordingly total mothers, knowledge were divided into:
   - Satisfactory knowledge, if score ≥ 60%.
   - Unsatisfactory knowledge, if score<60%.

2. **Mothers' reported practices assessment sheet:** it was developed by the researcher to assess mothers' reported practices related to measures to prevent injury and avoid fracture during clothing. Also, mothers’ reported practices related to their role in dental care of their children, first aid of fracture, management of pain and caring of cast.

   **2. Scoring system:**

   Mothers’ reported practices were checked and scored as one degree to the step which was done correctly" and zero to the step which was not done or done incorrectly. The total score of mothers reported practices was 43 point. All scores were summed up and accordingly, total mothers' reported practices was classified into:
Satisfactory level of mothers reported practices, if score ≥ 60%.
- Unsatisfactory level of mothers reported practices, if score <60%.

3. Quality of life scale for mothers having children suffering from OI (WHO, 1998): It was modified by the researcher and translated into the simple Arabic language to suit the nature of the study. The scale divided into physical health domain (seven items), psychological health domain (six items), social relations domain (three items) and environmental domain (eight items) [7].

The quality of life scale for mothers is a four point scale, the numerical values allotted to each response as following, The response options range from 1 degree (very dissatisfied/very poor) to 5 degree (very satisfied/very good).

3. Total scoring of the WHO Quality of Life Scale (WHOQOL):
- Score ≤ 60% referred to low QoL.
- Score between 60% < 75% referred to moderate QoL.
- Score ≥ 75% referred to high QoL.

4. Quality of life scale for children suffering from OI (Hill et al., 2014): It was used to assess quality of life for children suffering from OI [8]. It includes 39 items related to child being safe and careful (six items), child reduced function (eight items), child pain (six items), child fear (six items), child being different (six items) and independence of the child (seven items).

4. Scoring system:

The quality of life scale for children with OI is a five point likert scale, the numerical values allotted to each response as following, the response options range from Always =5 degree, Most of the time =4 degree, Sometimes =3 degree, Not much =2 degree and Never =1 degree.

Total scoring of quality of Life Scale for children as categorized as the following:
- Score <60% referred to low QoL.
- Score 60%: < 75% referred to moderate QoL.
- Score ≥75% referred to high QoL.

Data collection procedure:

The actual field work was carried out over a period of 14 months from beginning of January 2019 up to the end of February 2020. The researcher attended the study setting according to
The actual field work was divided into four phases:

**Assessment phase:**

In this phase, the researcher used the constructed tools for collecting the data about mothers' knowledge and their reported practices, as well as their quality of life related to care of their children suffering from OI (pre-test). The purpose of the study and its expectations were explained by the researcher to the studied mothers before starting data gathering.

**Planning phase:**

The guideline program intervention was designed on the light of the literature review and modified according to mothers’ educational needs regarding care of their children suffering from osteogenesis imperfecta.

**Implementation phase:**

The program was implemented in sessions theoretical and practical to each mother or in groups that ranged from 10-15 mothers according to their physical and mental readiness at the studying room in the genetic department of Children Hospitals affiliated to Ain Shams University Hospital and time of sessions conduction was organized according to telephone call to each subject.

**Evaluation phase:**

This phase included evaluation the effect of implementation of guideline program interventions on mothers having children suffering from osteogenesis imperfecta by comparing the results of pre & post test through used the same tools. Mothers and their children were evaluated after six months post guideline program intervention.

**Ethical consideration:**
Oral consent was obtained from each mother as a prerequisite to be engaged in the study prior to fill the study tools after clarification of the study aim, methods and possible outcomes of the research and assuring them that, the gathered information will be used for scientific research purpose only and will be strictly confidential. Study subjects were free to withdraw from the study any time they want. Each mother was secured and ensured that the study is harmless, confidentiality of the gathered data, voluntary participation, and they are free to withdraw quit.

**Statistical analysis:**

Data entered and manipulated through statistical package for social science and revised, coded, tabulated and presented using statistics in the form of frequencies and percentages. Mean and standard deviations were used for quantitative variables. The qualitative data were also analyzed by applying appropriate statistical method chi square ($x^2$) to determine whether there was a statistical significant difference or not. The statistical significant difference was considered as follows:

- Non-significant (NS) $p > 0.05$
- Significant (S) $p \leq 0.05$
- Highly significant (HS) $p \leq 0.01$

**III. RESULTS**

The mean age of studied mothers was 32.82±5.3 years and of the studied children was 8.14±5.71 years. Majority (91.3%) of studied mothers had negative family history of OI and less than two thirds (63.7%) of them had consanguineous partner.

More than three quarters (78.6%) of the studied mothers had satisfactory level of knowledge post program intervention compared to 42.5% of them pre program intervention ($x^2=20.132$ **P-value** $<0.001$), *figure (1).*

82.9% of mothers had adequate reported practices post program intervention compared to less than half (48.75%) pre-program intervention ($X^2=19.007$ **P-value** $<0.001$), *figure (2).*

Nearly three quarters (74.3%) of the studied mothers had high level of quality of life post program intervention compared to 22.5% of them pre-program intervention ($X^2=46.741$ **P-value** $<0.001$), *figure (3).*
As shown in figure (4), more than two thirds (67.5%) of studied children had high level of quality of life post program intervention compared to 38.6% pre program intervention ($X^2=7.900$, P-value <0.019*).

There was a positive correlation between all variables of the study namely total mothers’ knowledge, reported practices, QOL of both studied mother and their children post program intervention, table (1).

**Figure (1):** Total knowledge of the studied mothers regarding OI pre/post guideline program intervention.

**Figure (2):** Total reported practices of the studied mothers regarding to care of their children suffering from OI pre/post guideline program intervention.
Figure (3): Total quality of life of the studied mothers’ pre/post guideline program intervention.
**Figure (4):** Total quality of life of the studied children pre/post guideline program intervention.

<table>
<thead>
<tr>
<th>Items</th>
<th>Total knowledge</th>
<th>Total reported practices</th>
<th>Total QOL of mother</th>
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<tr>
<td></td>
<td>r</td>
<td>P-value</td>
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<tr>
<td>Total reported practices</td>
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<td>Pre (n=80)</td>
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<td>&lt;0.001*</td>
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<td>Post (n=70)</td>
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<td>Total QOL of children</td>
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<tr>
<td>Pre (n=80)</td>
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<td>&lt;0.001*</td>
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<tr>
<td>Post (n=70)</td>
<td>0.432</td>
<td>&lt;0.001*</td>
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*Significant p<0.05  ** Highly significant p<0.001

**Table (1):** Correlation between study variables pre/post guideline program intervention.

**IV. DISCUSSION**

The current study aimed to evaluate the effect of guideline program interventional on mothers of children with OI through assessing mothers' knowledge and practices regarding caring of their children, designing and implementing interventional program and evaluating the effect of this program on mothers' knowledge, practices and quality of life.
Concerning to the total knowledge of the studied mothers regarding OI pre/post guideline program intervention, the results of present study showed that, more than two fifth of them had satisfactory knowledge regarding OI pre program intervention. From the researcher point of view, unsatisfactory level of the studied mothers’ knowledge pre guideline program intervention might be due to inadequate information given by healthcare team and the nature of the disease which is considered a rare disease. This result in the same line with Castro et al., (2016) who mentioned that, family caregivers of children with OI face unique challenges in providing care, which include limited access to information about the condition and ongoing care[9]. Also, reported many unknowns about the disease, how to proceed with treatment compared to more than two fifth of them pre guideline program intervention and this reflected statistical significant difference between pre and post guideline program intervention. This result in the same line with Dogba et al., (2016) who mentioned that, most of the studied mothers had satisfactory knowledge regarding OI post program intervention compared to [10].

As regards the studied mothers’ total reported practices regarding to care of their children suffering from OI pre/post guideline program intervention, the present study showed that, less than half of them had adequate reported practices regarding to care of their children suffering from OI pre guideline program intervention. From the researcher point of view, this result may be due to the guideline program intervention improved the mothers total level of reported practices regarding to care of their children suffering from OI. This result was congruent with Collins et al., (2016) who found that, caregivers of children with OI became experts in caring for their children's unique daily health and care needs, developed strategies which make future day-to-day caregiving work easier and learned to manage the environment to be safer, coordinating with schools to manage the care of their child safely and searching for low-impact activities for their children [11]. While, most of the studied mothers had adequate reported total practices regarding to care of their children suffering from OI post guideline program intervention. This result was in the same line with Castro et al., (2020) who illustrated that, most of the studied mothers had adequate reported practices regarding to care of their children suffering from OI [12].
Concerning the studied mothers’ total quality of life pre/post guideline program intervention, the results showed that, one quarter of the studied mothers had lowest quality of life related to physical, psychological, social and environmental domain pre guideline program intervention. From the researcher point of view, this result due to the dependency of children on their caregivers during their care, lack of support from their relatives, mothers fear from dependent on others in caring of their children and fear from having other children with the same disease pre guideline program however, post guideline program the mothers had adequate level of knowledge and practices that enable them to be more knowledgeable to care for children with OI. This result was in similarity with Arabaci et al., (2015) who mentioned that, caregivers of children with OI had burned out, hopeless and overwhelmed by the unique challenges of caring for children with brittle bones and reported lower scores in the environmental quality of life domain as environmental safety and access to social supports. Compared to more than two thirds of them had high quality of life related to physical, psychological, social and environmental domain post guideline program intervention. This result was incongruent with Vanz et al., (2018) who mentioned that, the mothers of children with OI had lowest quality of life (QOL) for physical domain, psychological domain, social domain and environmental domain post intervention. Also, this result was disagreed with Hill, Baird and Walters. (2016) who illustrated that most of the studied mothers had high quality of life post program intervention compared to few number of them pre guideline program intervention.

According to the studied children total quality of life pre/post guideline program intervention, the current study results clarified that more than two thirds of the studied children had high quality of life post guideline program intervention compared to more than one third pre guideline program intervention. From the researcher point of view, this result might be due to over protective of mothers to their children during caring of them which lead to dependency of children on their mothers, fear from dealing with other even their sibling. Also, this result might reflects the nature of their disease which characterized by fragile bone and lead to recurrent fractures and this disease might effect on their psychological status (feeling that they different from other children) despite their young age. This result in similarity with Tsimicalis et al., (2018) who illustrated
that, more than two thirds of the studied children had high quality of life after the Zoledronate (bisphosphonate) infusion compared to less than one third pre the Zoledronate infusion [16].

Concerning the correlation between study variables pre/post guideline program intervention, the results showed that there was positive correlation between all variable of the study subject namely, total knowledge, total reported practices, total QoL of mother& total QoL of child. These results were congruent with a study done by Elsobky et al., (2018) who mentioned that, there was highly statistically significant difference in relation to mothers’ knowledge and reported practice in pre and post implementation of program intervention [17]. Also, these results were disagreed with Vanz et al., (2019) who mentioned that, there wasn’t correlation between caregivers’ total knowledge and their total QoL regarding to care of their children suffering from OI [18]. The present study results were in agreement with Castro et al., (2020) who mentioned that, there was positive correlation between total practices of mothers, total QOL of mothers and total QoL of child [12].

CONCLUSION

Based on results of the current study, it can be concluded that, there were significant improvements of mothers' knowledge and practices regarding to care of their children suffering from OI post implementation of guideline program intervention. Moreover, there were positive correlations between all variables of the study namely total knowledge, total reported practices, total QOL of mother and total QOL of child. Based on the present study finding it is suggested that further researches should be conducted to determine the barriers/ challenges that affect negatively on the mothers in caring of their children with OI.

V. ACKNOWLEDGEMENTS

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Ethical Clearance: Institutional ethical committee obtained for the study.
Source of Funding: self
Conflict of Interest: Nil
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