






ORIGINAL ARTICLE

NURSE'S CHALLENGES REGARDING CHILDREN WITH OSTEOGENESIS IMPERFECTA*

DESAFIOS DOS ENFERMEIROS FRENTE À CRIANÇA COM OSTEOGÊNESE IMPERFEITA

DESAFÍOS DE LOS ENFERMEROS EN EL CUIDADO DE NIÑOS CON OSTEOGENESIS IMPERFECTA

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ABSTRACT






Objective: to identify nurses' challenges in the care of children with osteogenesis imperfecta. **Method:** this is a qualitative, descriptive and exploratory study with 14 on duty nurses in the pediatric complex, who answered a questionnaire. Data was analyzed by Content Analysis. **Results:** it was understood that the care of these children becomes a challenge, since any inappropriate intervention related to mobilization may trigger complications due to bone fragility. **Conclusion:** it is pointed out that the study served as learning for a disease that is not well known, which instigates some health professionals regarding its management. In this context, the systematization of nursing care for patients with OI becomes indispensable, as they require special care. **Descriptors:** Nursing; Orthopedics; Delivery of Health Care; Child Care; Nurses Pediatric; Osteogenesis.

RESUMO

Objetivo: identificar os desafios dos enfermeiros na assistência à criança com osteogênese imperfeita. **Método:** trata-se de um estudo qualitativo, descritivo e exploratório, com 14 enfermeiras plantonistas do complexo pediátrico, que responderam a um questionário. Analisaram-se os dados pela Análise de Conteúdo. **Resultados:** entendeu-se que o cuidado dessas crianças se torna um desafio, visto que qualquer intervenção inapropriada relacionada à mobilização poderá desencadear complicações devido à fragilidade óssea. **Conclusão:** aponta-se que o estudo serviu como aprendizado para uma doença não muito conhecida, a qual instiga alguns profissionais de saúde em relação ao seu manejo. Torna-se indispensável, nesse contexto, a sistematização da assistência de Enfermagem aos portadores de OI, já que necessitam de cuidados diferenciados. **Descritores:** Enfermagem; Ortopedia; Assistência à Saúde; Cuidado da Criança; Enfermeiras Pediátricas; Osteogênese Imperfeita.

RESUMEN

Objetivo: identificar los desafíos de los enfermeros en el cuidado de niños con osteogénesis imperfecta. **Método:** este es un estudio cualitativo, descriptivo y exploratorio con 14 enfermeras pediátricas en el complejo pediátrico, que respondieron un cuestionario. Los datos fueron analizados por el Análisis de Contenido. **Resultados:** se entendió que el cuidado de estos niños se convierte en un desafío, ya que cualquier intervención inapropiada relacionada con la movilización puede desencadenar complicaciones debido a la fragilidad ósea. **Conclusión:** se señala que el estudio sirvió como aprendizaje para una enfermedad no conocida, lo que instiga a algunos profesionales de la salud a tratarla. En este contexto, la sistematización de la atención de enfermería para pacientes con OI se vuelve indispensable, ya que requieren atención diferenciada. **Descriptores:** Enfermería; Ortopedia; Prestación de Atención de Salud; Cuidado del Niño; Enfermeras Pediátricas; Osteogénesis.

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INTRODUCTION

Osteogenesis imperfecta (OI) is known to be a disease characterized by bone fragility triggered by a qualitative or quantitative defect of collagen type one synthesized by osteoblasts.¹ The frequency of 6-7 / 100,000 individuals has been shown in studies, and in Brazil it is estimated that there are around 12,000 individuals with this diagnosis.²

OI is defined as a rare genetic disease that primarily affects tissue rich in collagen type one present in bone tissue, ligaments, tendons, dentin, sclera and skin.³ The main characteristic is bone fragility, caused by the decrease in bone mass, a condition that favors the appearance of bone deformities due to recurrent fractures.⁴

The so-called diagnostic triad, established by bluish sclera, dentinogenesis imperfecta, and generalized osteoporosis, is commonly used in a patient with multiple fractures or curving of long bones. The disease is also known by the terms "brittle bone disease" and "glass bone syndrome".⁵

Studies have reported that OI has no difference in gender or ethnicity distribution. The worldwide incidence of OI is estimated at 1 / 10,000- 20,000 live births.⁶

It is understood that OI, as a rare disease, does not have wide wisdom and management by health professionals, however, individuals affected by such pathology need specialized assistance for their clinical condition.^{1,4}

For the definition of the research scenario, a survey was conducted from the institute's databases and it was found that 164 children were hospitalized from 2009 to 2019 with diagnosis of OI in order to undergo some surgical treatment due to injuries caused by the underlying pathology. Among the children who underwent hospitalization and surgery, 7.4% were infants, 27% were preschoolers, 45% were school age and 20.6% were adolescents.

Given the above, it is understood the importance of a qualified professional to meet the demand of patients affected by OI.

It is argued that the diagnosis should be considered and investigated in any child with recurrent fractures, family history, clinical examination and radiological findings for diagnostic confirmation. It is also a laboratory diagnosis in the evaluation of calcium metabolism, and is useful for ruling out pre-existing hypokalemia and hyperparathyroidism.¹

In a relatively recent past, OI treatment was conducted by conservative measures, which stimulated minimal physical activity and, when the fracture was installed, was treated with surgical correction. Currently, treatment is associated with surgical-orthopedic procedures for

the treatment of fractures or deformities together with drugs that reduce bone fragility.⁷

It is pointed out that several treatments have been used in an attempt to obtain increased bone strength of patients with OI, but there is not yet a drug that has a specific action in this regard. The use of pamidronate, a bisphosphonate class substance, effective in children and adolescents with OI, has been shown to be associated with improved bone mass and decreased number of fractures.⁸

Bisphosphonates are substances that inhibit bone resorption, contributing to the efficiency of surgical treatment, increasing bone mineral density and providing a better quality of life for individuals with OI.²

As a requirement for bisphosphonate use, the presence of moderate or severe phenotypes, types one, three and four, is considered to be the first associated with chronic pain. It is recommended to use it under the following conditions: more than three fractures per year, the last two years without significant trauma; need for surgery due to vertebral fractures or deformities and radiographs showing signs of OI.¹

Patients with osteogenesis imperfecta are known to need care from their support network and all family support in an attempt to avoid fractures. This requires both home and recreational environments to have a different architecture to minimize the risk of fractures. It is noteworthy that the use of orthoses and pneumatic immobilizations can improve the quality of life of these patients, together with regular low-impact exercise and physical therapy to strengthen muscles, which consequently increase bone density.⁸

On the other hand, it is emphasized that the most effective surgical method in the treatment of disease manifestations is based on the work of Sofield and Millar. It is established that the method consists of multiple osteotomies, fragment realignment and spinal cord fixation for long bones, providing stability to the bones. The postoperative cast is immobilized until osteotomies are consolidated.⁵

Trauma-Orthopedic Nursing is defined as a specialized area related to assistance in disease situations, congenital processes and development, trauma, metabolic disorders, degenerative diseases, infections, and other impairments affecting the musculoskeletal, joint, and supporting connective tissue. It is emphasized that the specialty covers prevention, care and rehabilitation of individuals and, as most orthopedic diseases have long-term development, nurses must be able to identify problems and implement interventions early.⁶

It becomes essential, given the implications and challenges imposed on the management of children with OI for health professionals and especially for Nursing, care provided be guided by the principles of bioethics, which values care that maximizes benefits and minimizes harm to the patient.⁷

It is known that the Nursing process represents a systematic and humanized method of care consisting of five phases: research; nursing diagnosis; planning; Nursing implementation or interventions and evaluation.²

Thus, the nurse's work process leads her to research what she is doing and to evaluate how she could do it better, allowing the use of clinical reasoning to improve the quality of patient care. Thus, the constant evaluation and the probable readjustments of nursing actions and planning are possible.

It is noted that infants and children with OI require careful handling to avoid fractures. Support them when positioned, turned, moved or placed on their lap. It is emphasized that even during a diaper change, fractures can be caused in children with the severe type of the disease. Please be advised that these children cannot be lifted by the ankles to change the diaper; they should be lifted by the buttocks or placed on a pillow. In addition, occupational planning and genetic counseling are part of the long-term care goal.⁹

OBJECTIVE

- To identify nurses' challenges in caring for children with osteogenesis imperfecta.

METHOD

It is a qualitative, descriptive and exploratory study. The research scenario was defined as the pediatric complex of a federal public hospital located in the city of Rio de Janeiro (RJ), a reference in Traumatology and Orthopedics, the Pediatric Orthopedics of the hospital is a broad specialty that includes the treatment of diseases and birth defects, developmental disorders and the treatment of fractures that affect infants, children and adolescents, with 24 beds in their ward and four beds in the children's intensive care center.

The participants of the study were the nurses on duty in the pediatric complex, who signed the Free and Informed Consent Term (FICT) before the questionnaire.

The research included nurses on duty in the pediatric complex and excluded routine nurses, the head of the pediatric team, the nurses who work in the Additional Hospital Duty (AHD), those who work in the administrative area, are on sick

or vacation leave and have refused to sign the FICT.

The research sample was calculated according to the finite population formula. This calculation was based on a sample error of 5% and a confidence level of 90%, thus reaching the result of 17 nurses, corresponding to 100% of respondents, in order to guarantee the desired confidence in the research.

After approval by the Ethics Committee, nurses who met the established criteria participated in the study. The study was conducted in 2019, with data collection beginning upon project approval at Plataforma Brasil. The study participants were invited and, after clarifying the objectives and procedures, they were asked to participate by signing the FICT, following the ordinances of Resolutions 466/12, 510/16 and 580/2018 of the Ministry of Health, it should be noted that if for any reason they wished to abandon the study, they would be free to do so at any time and, after acceptance, the questionnaire was applied.

This project was submitted and approved by the Research Ethics Committee of the proposing institution for the initiation of the study under CAAE Number: 14907519.0.0000.5285 and Opinion 3.457.177 and from the Co-Participating Institution under CAAE Number: 14907519.0.3001.5273 and Opinion 3.502.866 entitled "Challenges faced by nurses in the care of children with osteogenesis imperfecta".

For the interpretation of the research data, the method of Content Analysis was used. Using content analysis presupposes three important steps: pre-analysis; material exploration and treatment of results and inference and interpretation. It is pointed out that the first phase consists of every process that will allow the work to be outlined and should be objective, with well-defined procedures. A fluctuating reading of all data is performed, that is, a first contact with the studies that will be subjected to the analysis, their selection, the creation of hypotheses and goals, the creation of the indicators that will guide the interpretation and the formal preparation of material. In the second stage, the coding units are chosen and, in the third and last stage, the integration of the categories into larger themes is performed.

RESULTS

According to the National Health Council (NHC), in order to guarantee anonymity and prevent the breach of information confidentiality, all respondents, at this stage of the research, were identified by country names related to the order of their interviews.

8 0 Table 1 shows the profile of the research participants investigated according to gender, age group, marital status, number of children, length

of professional experience and contractual relationship.

Table 1. Profile characterization of research participants. Rio de Janeiro (RJ), Brazil, 2019.

Variables	n	%
Female	14	100%
Age group		
25 to 30 years	1	7%
31 to 36 years	3	21%
37 to 42 years	8	58%
Over 42 years	2	14%
Marital status		
Single	6	43%
Married	6	43%
Divorced	2	14%
Number of children		
None	4	28,5%
One	6	43%
Two	4	28.5%
Experience time		
Up to five years	1	7%
Six to 11 years	-	-
12 to 17 years	9	64.5%
More than 17 years	4	28.5%
Contract bond		
Tenure	12	86%
Hired	2	14%

The pediatric complex is composed of 18 nurses, and one nurse was unable to participate in the research because she was the study's coordinator, two other nurses were on vacation and one nurse was on sick leave. It is noted that the 14 (100%) study participants were women. Historically, there is a predominance of women in the Nursing profession, since care is socially linked to the image of women.¹⁰

Regarding age, most professionals were aged between 37 and 42 years old (58%). Thus, it is evaluated that the age range of nursing professionals varies from 30 to 40 years, therefore, ages characterized by productivity,

which certainly ensures openness to knowledge of high complexity, enhancements and specializations.⁷

Most of the interviewees had professional qualification in more than one area, having as experience time between 12 and 17 years (64.5%). These findings lead to the understanding that it is the younger nurses who seek professional experience. It is also demonstrated that the professionals sought to improve their knowledge regarding the exercise of their profession, suggesting that they have property when talking about the subject, such a transversal and important theme in care.¹¹

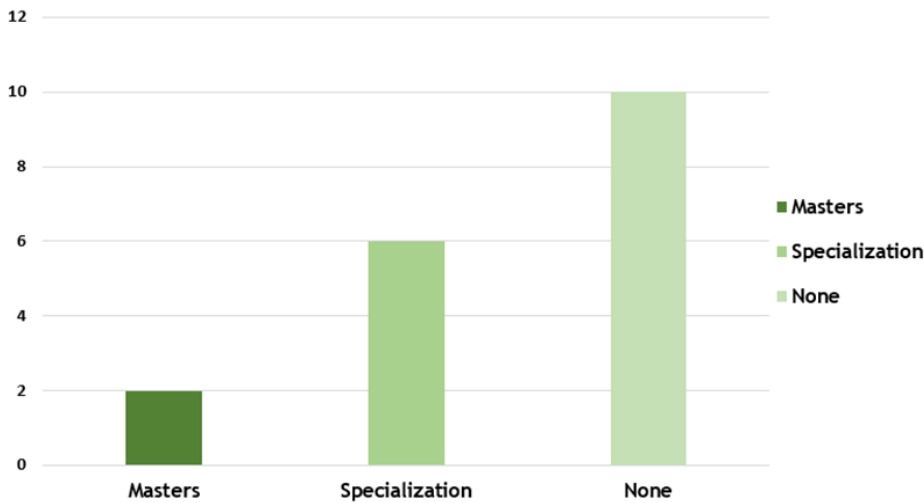


Figure 1. Post-graduation. Rio de Janeiro (RJ), Brazil, 2019.

Based on the specific curricular guidelines for the Nursing course, the objective is to train a generalist nurse capable of recognizing the most prevalent health-disease problems / situations in the national epidemiological profile and intervening in them. The professional, through the

Lato sensu postgraduate courses in Nursing, proposes a preparation directed to a determined area of knowledge in order to improve the practice of patient, user, family and community care.⁶

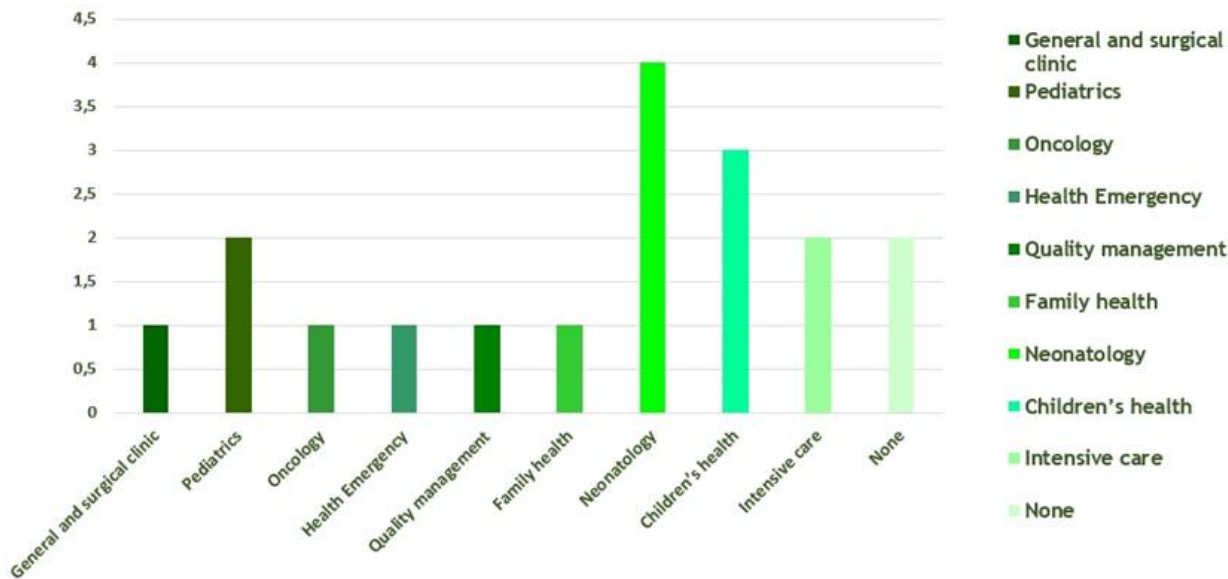


Figure 2. Area of specialization. Rio de Janeiro (RJ), Brazil, 2019.

In the case of specialization, two nurses had, besides the title of specialist, the masters. It is noted that the production of care has been based on the experience of nurses and the individual search for knowledge, demonstrating the difficulties faced by undergraduate nursing students and the importance of professional training.¹⁰

DISCUSSION

It follows from the materiality that emerged from the nurses' speeches, the Content Analysis according to Bardin. After characterization of the research participants, the analysis of the interviews.

♦ Category 1: Care in assistance due to handling and mobilization

It is noteworthy that procedures involving the movement and transportation of patients are considered the most painful and dangerous for health workers. Careful planning is required before beginning these procedures. In this context, basic guidelines and procedures were developed that had theoretical support in the international literature.¹²

As fractures are the main problem of children with OI, it is evaluated that the care of these children becomes a challenge for caregivers, since any inappropriate intervention related to mobilization may trigger complications due to bone fragility. False accusations of child abuse are considered common to family members of children suffering from mild forms of OI and / or those in whom the disease has not yet been diagnosed. Typically, some types of fractures are observed in both situations: fractures at different stages of healing; rib fractures; of spine and fractures that do not match the cause described, which ends up victimizing the family.¹³ Individuals severely affected by OI are known to require wheelchair use for community mobility, but may acquire transfer skills and personal care. It is often found

that they may need psychological support related to their body image.⁶

It is understood that to overcome the issues of impaired physical mobility, the caregiver needs help, which often limits the outings with the child, isolates them from their family world and, consequently, deprives them of relationships with the social environment.¹⁴

It is pointed out that the systematization of nursing care is extremely necessary, even in nurseries where OI patients are in need of special care due to their fragility. It is pointed out that “it is up to Nursing to seek knowledge, improve it and to plan systematic actions aimed at achieving a level of quality compatible with the needs of the child and their family”. It is argued that Nursing should encourage parents to participate in child care in order to teach them how to bath, change diapers and dress so that new fractures can be avoided.⁸

Many individuals with genetic diseases are known to live with the conditions imposed by a chronic disease, such as OI, in which the individual needs daily care to avoid fractures and treatment that requires periodic consultations, corrective surgery, in addition to the use of medications and physical therapy. Fractures are highlighted as the most common injuries in children and adolescents with OI, who, when they realize the need for immobilization, suffer much discomfort and limitations.¹⁵

It is considered that the nurse's role in caring for a child or newborn with OI is to provide safe care with a focus on preventing new fractures and worsening existing fractures, as well as a constant assessment of the child so that they can be recognized, if they occur. It is essential to know the classic signs of a fracture: heat; edema; hyperemia and discoloration at the site of injury and sometimes fever. It is observed that the actions should provide pain relief, so the position change should be performed with extreme regularity and the child's handling should be done

in order to avoid the risk of fractures. It is argued that professionals should also pay attention to the positioning of the child in bed, in order to prevent respiratory impairment.¹⁶

At the present time, the treatment that contributed most to improve patients' quality of life is bisphosphonates, drugs that improve muscle strength, help mobility, increase vertebral bone mass, decrease bone pain and the number of fractures, helping to increase the possibilities of surgical treatment of deformities.¹⁷

It is known that in adolescence (13-18 years), young people usually go through a troubled period in hormonal, social, emotional and relational levels. In particular, young people with osteogenesis imperfecta who use wheelchairs are concerned about mobility, social problems related to short stature and pain. It is observed that the adolescence phase can also be characterized by the existence of recurrent fractures. Regarding the social domain, it is noticed that young people are more concerned with appearance, since they have short stature and structural deformities due to previous fractures. It is known that in a young person with OI, there are often feelings of inadequacy and depression caused by the visibility of their limitations. In addition, they often wear orthopedic appliances, wheelchairs or other devices without forgetting their short stature.¹²

The studies showed the feelings of caregivers of children and adolescents with OI at diagnosis, highlighting that 93.5% were mothers. All caregivers reported that they felt anxious (100%), while 44 (95.7%) felt sad, 41 (89.1%), puzzled, 40 (87.0%), nervous, 40 (87.0%), frightened, 39 (84.8%), disappointed, 29 (63%), shocked and 28 (60.9%), depressed. It is shown in this study that the caregiver presents a great tension related to care.⁴

Hospitalization is considered a very common situation, but it generates conflicting feelings and feelings for the child, as it removes them from their daily life, from the family environment and promotes a confrontation with pain, limitation and passivity, causing the feeling of guilt, punishment and fear of death. It is necessary, to overcome this experience, that the child has instruments of his domain and knowledge.¹⁴

It is considered that a health establishment that ensures the most humanized conditions of care and physical structure in the inpatient units is the one that best serves its clientele, promoting the humanization of the hospital environment and collaborating with the patient's therapeutic process, contributing to the quality of health services provided by the professionals involved.⁹

♦ Category 2: Immediate postoperative care and with surgical wound

Nursing care is started for surgical patients at the time of admission. It is argued that it is up to the nurse to worry about making the hospital environment as receptive as possible. It is known that the surgical act is part of a situation involving several stressors and high tension generators that begin with the patient's arrival at the hospital. It is pointed out that anxiety is a common condition, presented by patients in the preoperative period, and the nurse should be alert to the signs of this situation to help them. The establishment of a trusting relationship between the nurse and the patient is seen as fundamental for them to be able to express their feelings.¹⁸

In addition to all the functions, it is verified that the nurse is also responsible for identifying the pain-related aspects of postoperative patients, and should have knowledge of theories and concepts on pain, anatomy and physiology of pain pathways, on pain relief and measurement methods, pharmacological management and their possible adverse reactions, as well as non-pharmacological care and prevention of complications.¹⁵

Nursing prescription is adopted as a means to improve the efficiency of services, being characterized as a service in which the demand goes beyond resources, aiming at improving postoperative patient care. It is necessary to record vital signs at standard times, to observe pain faces, to register the acceptance of the diet, to register the vesicointestinal eliminations, to keep the bed grid elevated and to stimulate playful activities.¹¹

It is also necessary to observe the surgical dressing for bleeding, observe and record the perfusion, temperature and coloration of the child's extremities in order to investigate the circulation and / or compression, observe compressive signs exerted by the plaster, perform body hygiene once a day, guide the companion on postoperative care, observe the sleep and rest pattern and offer a peaceful environment.¹⁹

In the postoperative period, regardless of the specificity of the surgery, general care is performed, such as the patient's transfer from the stretcher to the ward bed, thermal control, maintenance of respiratory function, observation of the general condition, the verification of dressing abnormalities, observation of the functioning of probes, the control of vital signs, the promotion of comfort and safety through the appropriate environment, the change of position, the promotion of active and passive movement, ambulation and family orientation for discharge. The importance of individualizing orthopedic patient care is emphasized, taking care not to fall into the routine sequence of general care to surgical patients.⁴

Among the specific care, those related to the plaster cast, orthopedic appliance and ambulation stand out. It is known that postoperative care aims at better rehabilitation of the surgical patient. It is observed that the assistance is differentiated, given the specificity of the place where the procedure was performed. Moreover, it is understood with technological advances in the orthopedic area, that the use of implantable devices has minimized patient discomfort, improving the quality of life in relation to the mobility deficit.⁹

♦ **Category 3: Permanent education**

The Ministry of Health (2004), through the Secretariat of Labor Management and Health Education, established the National Policy on Permanent Health Education, defined as learning at work where learning and teaching are incorporated into the daily life of organizations. It is a strategic tool for the qualification of workers, which allows a change in the conception and training practices, making it possible to add learning to the daily life of the organization, valuing the practice as a source of knowledge and actively inserting the professional in the educational process. In addition, the interaction of the interdisciplinary team is promoted and discussion and educational spaces are expanded.¹¹

The perspective of permanent health education for the nursing team is considered of paramount importance, since it acts as a protagonist in the construction and maintenance of health care. Thus, it is understood that the team needs to be able to meet the demand presented by the assisted clientele.¹⁷

It is necessary, as it is a scenario where care has as its central object the child with osteogenesis imperfecta and his family, to emphasize the discussion, the knowledge and the pathophysiological and semiological peculiarities, besides the complications, in order to allow nurses to build an evidence-based praxis, to implement actions, propose interventions and systematize appropriate care for this type of patient, thus enabling greater comfort, safety and improvement in their quality of life, since the disease in question has no cure and the possible therapeutic approaches are palliative, focusing on the permanent education of the patient and their families and on the prevention of new fractures and reduction of bone deformities.¹³

It is affirmed, confirming the report above, that “a prepared nursing is capable of preventing positional contractures and deformities, providing pain relief, handling the child so that the risk of fractures is not so imminent”.⁸

It is noteworthy that the care for the individual with OI covers from birth to adulthood, and nursing is present throughout this trajectory, and

it is essential to have knowledge of such pathology to promote quality and effective care.¹⁶

It is pointed out that children with OI should be assisted by a specialized care network, through a multidisciplinary team, in order to meet the needs imposed by the pathological condition. Likewise, it is argued that the multidisciplinary team should be qualified to provide, in addition to providing specialized assistance, tools to empower parents / family members in caring for the child.¹⁷

♦ **Category 4: Creating OI Patient-Specific Protocols / Interface Routines**

The Standard Operating Procedure (SOP) is the basis for ensuring the standardization of an institution's tasks, guaranteeing its users a service or product free of undesirable variations in its final quality.¹²

Nursing has been using this tool in order to improve its processes, as it allows all workers to provide standardized care to the patient, in accordance with the technoscientific principles and also, contributes to resolving distortions acquired in practice and also has an educational purpose. Moreover, it is pointed out that the adoption of care protocols provides more satisfaction for the Nursing team, more security in performing the procedures and, consequently, more safety for the patient.¹⁵

It is noted that the adoption of systematized care, based on a standard operating protocol, can bring many benefits in care practice, especially with regard to pathologies such as OI, which have many peculiarities and specificities.¹⁹

CONCLUSION

Osteogenesis imperfecta is defined as a genetic disease with distinct clinical characteristics, such as bone fragility, recurrent fractures, bluish scleras and dentinogenesis imperfecta. It is important to know how to identify these characteristics to guide the diagnosis of nursing, optimizing treatment and differentiating the picture of other diseases that can also cause fractures.

In this study, the challenges faced by nurses in the care of children with OI were identified, describing the care provided and allowing to know the opinion of each nurse involved in the care process. It was noticed, when reflecting on the Nursing challenges applied to children with OI, that, although Nursing is aware of the importance of this type of care, she faces great difficulties in systematizing in face of this pathology and its complications.

It is noted that the lack of general and applied knowledge of the pathology, in addition to the rarity of the disease, makes it not properly addressed in undergraduate nursing courses. It is established, as another point that contributes

negatively to this scenario, the lack of publications, especially in Nursing, about how care should be directed to this child, promoting insecurity and unpreparedness to deal with this public, since there is no basic literature that can support the care.

It is concluded that the study served as learning for a disease not well known, which instigates some health professionals regarding its management. In this context, the systematization of nursing care for patients with OI becomes indispensable, as they require differentiated care.

Therefore, doors are opened for a series of discussions in the field of OI, and it is expected to contribute positively on the awareness of the importance of this theme, especially for Nursing professionals, who are responsible for the production and management of care.

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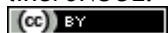
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