

# Nursing care for children and adolescents with epidermolysis bullosa: a systematic review

Cuidados de enfermagem com crianças e adolescentes com epidermólise bolhosa: revisão sistemática  
Cuidados de enfermería a niños y adolescente con epidermólisis ampollosa: revisión sistemática

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## Abstract

**Objective:** To analyze the scientific production regarding actions/Nursing Interventions in hospital environments related to the care of children and adolescents with epidermolysis bullosa.

**Methods:** This is a systematic review, which was searched in the CINAHL, MEDLINE®/PubMed®, Scopus, LILACS and SciELO databases, carried out from September 2020 to January 2021. For the search, the descriptors “epidermólise bolhosa” AND “criança” AND “adolescente” AND “enfermagem” were used, in Portuguese, in the LILACS and SciELO databases, and “epidermolysis bullosa” AND “children” AND “adolescent” AND “nursing” in the other databases.

**Results:** There was a greater number of articles published based on the guiding question having the United States as the country of origin (22%). Most of the classification was at level VI (44%) of scientific evidence. Still, 86% of studies involved research for the care plan. The evidence found resulted from expert opinions, case studies and consensus. The most cited care factors were skin care plans, diaper changing, clothing care and non-stick coating use.

**Conclusion:** The surveys reported difficulties regarding the availability of materials, treatment and specialized professionals, in addition to limitations of knowledge in clinical practice focused on the characteristics of epidermolysis bullosa. Among the care, there was emphasis on information about the wound complexity and characteristics as a way of anticipating care strategies.

## Resumo

**Objetivo:** Analisar a produção científica referente às ações/Intervenções de Enfermagem no ambiente hospitalar relacionadas ao cuidado com crianças e adolescentes com epidermólise bolhosa.

**Métodos:** Revisão sistemática, cuja busca se deu nas bases Cinahl, MEDLINE®/PubMed®, SCOPUS, LILACS e SciELO, realizada no período de setembro de 2020 a janeiro de 2021. Para a busca, foram utilizados os descritores “epidermólise bolhosa” AND “criança” AND “adolescente” AND “enfermagem”, nas bases Lilacs e SciELO, e “epidermolysis bullosa” AND “children” AND “adolescent” AND, “nursing” nas demais bases em inglês.

**Resultados:** Houve maior registro de artigos publicados com base na pergunta norteadora tendo como país de origem os Estados Unidos (22%). A maioria da classificação era no nível VI (44%) da evidência científica. Ainda, 86% dos estudos envolveram pesquisas para o plano de cuidados. As evidências encontradas decorreram de opiniões de especialistas, estudos de casos e consenso. Os fatores de cuidados mais citados foram planos de cuidados voltados à pele; troca de fraldas; cuidados com as roupas e uso de coberturas antiaderentes.

**Conclusão:** As pesquisas reportaram dificuldades quanto à disponibilidade de materiais, tratamento e

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profissionais especializados, além das limitações dos conhecimentos na prática clínica voltada às características da epidermólise bolhosa. Dentre os cuidados, houve destaque para informação sobre a complexidade e as características da ferida como forma de antecipar as estratégias de cuidado.

## Resumen

**Objetivo:** Analizar la producción científica referente a las acciones/intervenciones de enfermería en el ambiente hospitalario relacionadas con el cuidado a niños y adolescentes con epidermolísis ampollosa.

**Métodos:** Revisión sistemática, cuya búsqueda se realizó en las bases Cinahl, MEDLINE®/PubMed®, SCOPUS, LILACS y SciELO, realizada en el período de septiembre de 2020 a enero de 2021. Para la búsqueda se utilizaron los descriptores “epidermolísis ampollosa” AND “niño” AND “adolescente” AND “enfermería”, en las bases Lilacs y SciELO, y “epidermolysis bullosa” AND “children” AND “adolescent” AND, “nursing” en las demás bases en inglés.

**Resultados:** Con base en la pregunta orientadora, hubo un mayor registro de artículos publicados que tenían como país de origen Estados Unidos (22 %). La mayoría de la clasificación era de nivel VI (44 %) de la evidencia científica. Además, el 86 % de los estudios incluyeron investigaciones en el plano de los cuidados. Las evidencias encontradas derivaban de opiniones de especialistas, estudios de casos y consenso. Los factores de cuidados más citados fueron planos de cuidados orientados a la piel, cambio de pañales, cuidados con la ropa y uso de coberturas antiadherentes.

**Conclusión:** Las investigaciones indicaron dificultades en cuanto a la disponibilidad de material, tratamiento y profesionales especializados, además de las limitaciones de conocimientos en la práctica clínica orientada hacia las características de la epidermolísis ampollosa. Entre los cuidados, se destacó la información sobre la complejidad y las características de la herida como forma de anticipar las estrategias de cuidado.

## Introduction

“Epidermolysis bullosa” is the name given to a group of genodermatoses, being clinically classified into four genetically different subtypes: hereditary, non-contagious, rare and chronic and without cure. Its characteristics are formation of blisters, mutation in epithelial structural proteins causing fragility to the skin and ruptures, with great genetic variability.<sup>(1,2)</sup> Considering these characteristics, there are approximately 500,000 patients with epidermolysis bullosa worldwide, mostly children.<sup>(3)</sup>

According to estimates by the Dystrophic Epidermolysis Bullosa Research Association of America (Debra of America), the incidence of epidermolysis bullosa is approximately 19.57 cases per million live births.<sup>(4)</sup> In Brazil, it is estimated that there are approximately 1,600 people with epidermolysis bullosa, mostly children and adolescents.<sup>(1)</sup>

Despite the significant relevance of this problem, care management for epidermolysis bullosa is a challenge for nursing care, due to the complexity and variety of its manifestations. Furthermore, skin immaturity of newborns with epidermolysis bullosa requires specialized care, as its fragility causes the skin to rupture, with the formation of blisters at the slightest friction. The epidermis breaks down due to the spontaneous increase in temperature, causing blisters that can vary from mild to severe, depending on the subtype of epidermolysis bullosa.<sup>(5)</sup>

Healthy skin contains a layer of proteins, called collagen, capable of keeping it intact and which is responsible for uniting the cells of the most superficial layer of the skin with the innermost layer, providing resistance and having a protective function. In people diagnosed with epidermolysis bullosa, this collagen is absent or altered, causing the skin to rupture and blister.<sup>(1,5,6)</sup>

Thus, the main characteristic of epidermolysis bullosa is the appearance of chronic skin injuries that, in general, are small, numerous, painful, uncomfortable and exudate. The more serious subtypes of epidermolysis bullosa cause physical and emotional suffering to children and adolescents. They are often interconnected with the injury’s cutaneous involvement and can injure the eyes, nose, oral mucosa, dentition, gastrointestinal and genitourinary tracts and the musculoskeletal system, in addition to causing metabolic imbalances, such as malnutrition and anemia.<sup>(2)</sup>

Therefore, children’s and adolescents’ quality of life is affected by epidermolysis bullosa, requiring integrated, specialized and individualized assistance in health care, especially by professional nurses.<sup>(6)</sup> There are great challenges in the face of Nursing Interventions (NI), such as care plan variability, individualized management and availability of specific products, which generally have a high cost for the family budget and hospitals, specific knowledge about the disease complexity and specialist wound care.<sup>(7,8)</sup>

Nurses have relevant conduct, since they participate fully in the health care of these patients, whether for pain control and relief; observation of clinical signs; handling; performing dressings; change of position and reduction of pruritus, as well as guidance on care for the injury; nutrition and prevention of complications, with nursing care planning and instruction in care for families/caregivers.<sup>(9)</sup>

Caring for these children and adolescents in a pandemic period must be cautious, redoubling prevention and protection measures, in view of their susceptibility to injuries. Research has shown that severe acute respiratory syndrome coronavirus 2 (SARS-CoV-2) enters host cells using the spike protein to bind to the angiotensin-converting enzyme (ACE) cell receptor, which is manifested in skin capillary, in the basal layer of keratinocytes, in the eccrine glands, in the mucosa and in the nasal epithelium, which increases the vulnerability of these children and adolescents to the virus.<sup>(10-12)</sup>

The pandemic scenario is challenging, both for families/children-adolescents and for nursing professionals, due to the great complexity of care, combined with the limitations that the pandemic has generated, such as isolation and difficulty in accessing hospitals, with a lack of holistic and specialized care.<sup>(13)</sup>

Thus, for this care to be based on solid foundations, it is essential to develop research with a view to seeking evidence, but there are still few studies in scientific literature and related to NI in the hospital environment aimed at caring for children and adolescents with epidermolysis bullosa.<sup>(14)</sup>

Thus, in order to clearly and explicitly provide evidence on nursing care for children and adolescents with epidermolysis bullosa, the following question emerged: what has been produced about actions/IEs in the hospital environment related to child care and adolescents with epidermolysis bullosa?

To answer this question, the objective was to analyze the scientific production regarding actions/NI in the hospital environment related to caring for children and adolescents with epidermolysis bullosa.

## Methods

This is a systematic literature review, based on the Preferred Reporting Items for Systematic reviews and Meta-Analyses (PRISMA).<sup>(15)</sup> To prepare the research question, we followed the JBI recommendations, using the PICo strategy, with P for population, I for intervention and Co for context, for the review question formulation, in which P corresponded to children and adolescents with epidermolysis bullosa, I, to action/NI and Co, to hospital environments. From this, the following question was formulated: What are actions/NI in caring for children and adolescents with epidermolysis bullosa during the hospitalization period? We used the Journal Portal of the Coordination for Higher Education Personal Improvement – Brazil (CAPES - *Coordenação de Aperfeiçoamento Pessoal de Nível Superior*) to access the following databases: Latin American and Caribbean Literature in Health Sciences (LILACS), Medical Literature Analysis and Retrieval System Online (MEDLINE), Cumulative Index to Nursing & Allied Health Literature (CINAHL), PubMed, Scientific Electronic Library Online (SciELO) and Scopus.

Article search was carried out between September 2020 and January 2021, followed by the other phases of the research. We used the descriptors indexed in Health Sciences Descriptors (DeCS) and Medical Subject Headings (MeSH) in Portuguese and English, combined using the Boolean operator AND: “*epidermolise bolhosa*” AND “*criança*” AND “*adolescente*” AND “*enfermagem*”, in LILACS and SciELO, and “epidermolysis bullosa” AND “children” AND “adolescent” AND “nursing”, in the other databases. For the selection of publications, we included complete articles, published from 2010 to 2020, in Portuguese, English and Spanish, that addressed the guiding question theme, with a description of actions/NI applicable during hospital care. Duplicate citations were removed manually. We excluded articles of reflection, reviews, monographs, theses, congress annals and incipient studies.

Articles were independently assessed by two reviewers, who read the title and abstract, following the inclusion criteria. Subsequently, selected articles

were read in full. In case of disagreement or doubt, a third reviewer was contacted for clarification.

An electronic spreadsheet was built by the main author, including year of publication, journal, author, article title, research design, country, main results and Level of Evidence. Subsequently, these items were discussed according to the basic human needs theoretical framework by Wanda de Aguiar Horta, considering the three levels of life (psychobiological, psychosocial and psychospiritual), in order to structure the needs affected in mother-child care. Regarding the Level of Evidence, a proposed classification system in seven levels,<sup>(16)</sup> described in Chart 1 was used for assessment.

**Chart 1.** Relationship of Levels of Evidence and their respective definitions

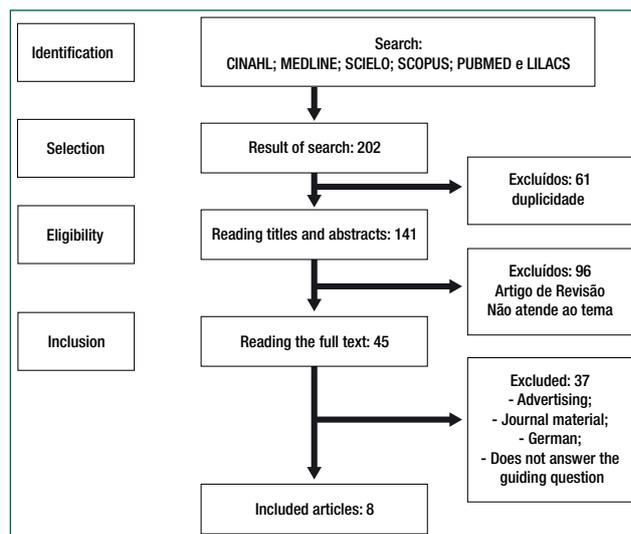
Level	Definition
I	Evidence from systematic review or meta-analysis of randomized controlled clinical trials; clinical guidelines based on systematic reviews or meta-analyses
II	Evidence derived from at least one well-designed randomized controlled clinical trial
III	Controlled clinical trial without randomization
IV	Case-control study or cohort
V	Systematic review of descriptive and qualitative studies
VI	Evidence derived from a single descriptive or qualitative study
VII	Opinion of authorities or report of expert committees

Source: Melnyk BM, Fineout-Overholt E. Evidence-based in nursing & healthcare: a guide to best practice. Philadelphia: Wolters Kluwer Health; 2015.<sup>(16)</sup>

## Results

Eight articles were analyzed. Figure 1 presents the synthesis of search results according to the databases consulted. The initial investigation in the databases using MeSH descriptors and terms resulted in 202 articles, with the following distribution: two from LILACS, seven from MEDLINE®, ten from CINAHL, 162 from Scopus, 21 from PubMed® and no articles from SciELO. Of these, when assessing the articles that met the inclusion criteria and the guiding question, eight studies remained, three (38%) from Scopus and five (63%) from PubMed®.

Of the 202 articles found during the search, only eight (4%) contemplated the guiding question. It is important to point out that 31 (15%) of them were unrelated to the guiding question of the study; 83 (41%) were not part of the theme; 61 (30%) were duplicates; 13 (6%) were integrative reviews; 1 (0%) was in another language; and 5 (2%) were advertise-



CINAHL: Cumulative Index to Nursing & Allied Health Literature; MEDLINE®: Medical Literature Analysis and Retrieval System Online; SciELO: Scientific Electronic Library Online; LILACS: Latin American and Caribbean Literature in Health Sciences

**Figure 1.** Flowchart of articles found in the databases searched

ments. Regarding the publication period from 2010 to 2020, oscillations were observed in publications in some years: 2010, 2013, 2016, 2017 and 2018 had no publications; there was 1 (13%) publication for the year 2011, 2012 and 2020 each; and 2014, 2015 and 2019 had two (22%) publications each. The studies had as countries of origin the United States (2; 22%), United Kingdom, Canada, Wales, France, Switzerland and Brazil (1; 13% for each) publication each. Regarding the classification based on Level of Evidence, it was identified that 4 (44%) belonged to level VI, 2 (22%) to VII and 2 (22%) to levels III and IV. Thus, 7 (86%) studies involved research for the care plan and/or educational instruction aimed at parents, nurses/health professionals and patients, in addition to skin care, such as changing dressings, care for clothes, hospital supplies, medications and specific products for epidermolysis bullosa. One (14%) was an observational study related to specific care for epidermolysis bullosa wounds, such as bathing with salt water. The evidence found stems from expert opinions, consensus, observational, retrospective study and studies and case reports. Since it is a rare disease, no experimental studies or methodologies were found for the development of new products and technologies to improve NI for children and adolescents with epidermolysis bullosa (Chart 2).

**Chart 2.** Scientific production on the subject

Authors	Design/Level of Evidence	Country/language	Main results
Secco et al. <sup>(14)</sup>	Case report/VI	Brazil/Portuguese	The following interventions were found: - In necrotic areas: simple coverage with sterile gauze (Rayon) soaked in essential fatty acids and hydrogel; - Analgesia at the time of dressing change: intermittent fentanyl, paracetamol and codeine; - Weekly change of dressings: covered with polyurethane foam and tubular mesh; - Guidance to parents/caregivers regarding care and changing dressings for injuries and the importance of changing positions every 4 hours.
Petersen et al. <sup>(17)</sup>	Observational, retrospective study/ III	United States/English	Salt water baths using the amount of ¼ to 2 cups of salt for a bathtub with a capacity of 35 liters of water (approximate concentration of 0.09% saline solution). There was a reduction in pain (discontinuity of analgesic use) and reduction of skin odor and skin secretion. Treatment may be recommended for all epidermolysis bullosa subtypes, as there was no statistically significant difference between the types of epidermolysis bullosa and all ages (children between 2 months and 13 years of age, research participants).
Packard et al. <sup>(18)</sup>	Case study/VI	United States/English	Educational instructions: - Avoid using adhesive material directly on the skin and/or that causes shear on the skin; - Choose using gauze with Vaseline; - Change dressings daily or as needed, preferably using Mepitel®; - Cleaning with topical liquid, such as ExSept Plus and Alcavis HDC diluted in saline.
Kearney et al. <sup>(19)</sup>	Inductive exploratory study/ III	Wales/English	Instructions directed to individual needs (patient and family): - Support the management of physical health issues (pain, itching, changing dressings and bathing); - Access to community/home services (geographical isolation of specialized services); - Specific information of epidermolysis bullosa and psychosocial support (absence of time for other activities and family breakdown); - Effective interaction with health professionals (nursing support); - Advice on benefits and rights (difficulty dealing with peers).
Denyer et al. <sup>(20)</sup>	Case study/VI	United Kingdom/English	Factors that may compromise the skin recovery process: suboptimal nutrition, anemia, pain, injuries and frequent blisters Material used: keratin gel providing epithelialization of the wound margins, causing a reduction in discomfort (change of dressings without analgesia with opioids); Gel use provided faster dressing changes, decreased heat, friction and itching.
Hachem et al. <sup>(21)</sup>	Multicenter consensus/ VII	France/English	Skin care: do not systematically place the newborn in an incubator and/or avoid heat; not using adhesive dressing; use thick padding before applying the cuff to measure blood pressure; put on clothes with few areas of seams; add silicone foam over diapers to reduce friction; regular bath, using soft felt towel; add protection to vulnerable skin spots (e.g., knees and elbows) with soft silicone contact layers; pay attention to friction during handling Wound care: the choice of dressings varies according to injury type and location, for exuding wounds, use hydrofiber and silicone foam dressings; in dry wounds, use non-adhesive and hydrogel soft silicone or hydrocolloid contact layers; Proper pain assessment and management are mandatory; Nutritional support started early; Vaccination schedule should be maintained.
Pope et al. <sup>(22)</sup>	Consensus study/ VII	Canada/English	A care plan for wounds was carried out with techniques for handling and caring for newborns, regarding: care with food; monitoring of hemoglobin levels; assessment of pain parameters; pruritus control for daily activities; foam dressing use (preferably those with padding for bony prominences); wearing clothes without elastics, diapers lined with non-adherent bandage or Barrier Cream or cream with zinc oxide; developing an assessment, location, and treatment plan for wound and pain
Schlueter et al. <sup>(23)</sup>	Case report/VI	Switzerland/English	Negative pressure wound treatment (NPWT), VAC® (KCI Medical®) with 75 mmHg suction, used continuously for 8 weeks with weekly dressing changes, was performed with a neonate. Initially, for wound site decontamination, hypochlorite solution (Veriforte®) was used. Hydrophilic polyurethane membrane dressings (PolyMem®) combined with an enzyme alginate (Flaminal Forte®). To fix the system, a transparent film dressing (Xtrata) was used, which was changed using an adhesive removal spray (Niltac™)

## Discussion

The results provide an overview of evidence on the main NI and point out that the theme still has little focus of interest for national and international scientific production. This analysis made it possible to identify the actions developed by nurses in hospital environments, in addition to the family's participation in this care process, associated with the complexity of knowledge and skills required in the care of children and adolescents with epidermolysis bullosa.

The actions carried out focus on aspects of care management, method of use and dressing tech-

niques and management of pain symptoms, in addition to interventions related to psychological and social impacts. Knowing that nursing practice must be guided by a theoretical basis,<sup>(24)</sup> Wanda Horta's theory of basic human needs was used to organize the evidence. This theory encompasses three levels of needs: psychobiological, psychosocial and psychospiritual.<sup>(25)</sup>

Psychobiological needs include challenging interventions for professionals and concern skin care, specifically blisters and injuries. In general, they arise spontaneously, are numerous, painful, uncomfortable and extensive.<sup>(19,20,22)</sup>

Understanding the entire structure of the disease with implications for severe subtypes and limitations on quality of life in the absence of specific and specialized interventions,<sup>(19,20,22)</sup> the main goals of treatment are to control or eliminate causal factors and provide targeted assistance to moderate or cease co-existing and potential agents by preventing secondary infections and promoting an optimal environment for wound healing. To this end, by consensus,<sup>(2,22)</sup> it is recommended to assess blister location and characteristics, such as a injury with a potential risk of infection, exudation and subtype of epidermolysis bullosa.

Two simple procedures, but considered highly risk factors for injuries, are dressing changes and friction during mobilization and changing clothes. It is usually a traumatic moment, and this leads to the appearance of new blisters and increased pain and itching, in addition to prolonging injury healing.<sup>(1,2)</sup> Aiming to reduce signs and symptoms, nursing appropriates the moment of bathing these patients to perform the atraumatic removal of coverings, using saline solution, in order to benefit children and/or adolescents in pain reduction, contributing to the improvement of wound healing and avoiding and/or reducing infections and itching.<sup>(17,22)</sup> However, it is suggested that studies with greater methodological rigor be developed, with all subtypes of epidermolysis bullosa: mild, moderate to severe.

Nurses and researchers support changing dressings during bathing with saline solution, considering the benefits already described, and that salt is an accessible and easily available solute, which can be acquired by family members. It is a non-invasive intervention that offers pain reduction, improves bathing adherence and decreases signs and symptoms of infection.<sup>(17)</sup>

The study pointed out that treatment with saline solution can be recommended for all subtypes of epidermolysis bullosa, between 2 months and 13 years, according to research participants.<sup>(17)</sup> It should be noted that, despite the limitations due to the sample, the results showed a decrease in secretion, odor, healing time, itching and analgesic use for bathing, with a reduction in pain.

Pain is a characteristic symptom of epidermolysis bullosa. Its frequency and intensity are usually proportional to the injury severity. Although multifactorial, injured skin significantly contributes to the symptom. With this, the nursing staff makes use of emollient and moisturizing products, keratin-based gel, non-adherent dressings, bath with warm and salty water, and instructs mothers, by encouraging exclusive breastfeeding or whenever possible, as breast milk favors the mother-child dyad, encourages safety and immunity and offers comfort to newborns.<sup>(17,21,22,26)</sup>

Patient's age is important in performing blister care. Infants with epidermolysis bullosa have extrauterine immunological immaturity and require immediate attention to avoid skin trauma.<sup>(14)</sup> Blistering in babies usually has a predisposition towards the extremities around the nappy area, and as age matures, babies become mobile, requiring soft knee pads and shoes to avoid shearing and thermal overheating.<sup>(21,26,27)</sup>

For blister care, NI should be started soon after birth, as parents' recognition of disease limitations of in daily life is important to overcome the challenges related to caring for a newborn who needs comprehensive and continuous care. For this, the presence of parents is necessary to strengthen the mother-child dyad and reduce family ruptures.<sup>(6,14,19,28,30)</sup>

From the perspective of non-drug interventions, there are a myriad of products that can be used, according to specificity and subtype of epidermolysis bullosa. The findings support the claim that foam dressing use is effective, as they contain a layer of silicone to reduce the appearance of new injuries and absorb exudate.<sup>(29,30)</sup> Dressings are designed to keep the wound clean, reduce chances of contamination, and promote healing, particularly of chronic wounds, which have significant potential for tissue loss.<sup>(14,21,30)</sup>

Pure hydrogel or associated with essential fatty acids contributes to autolytic debridement and protects the granulation tissue. Hydrofibers favor moisture at the injury site and reduce physical trauma and pain. Gauze with essential fatty acids can benefit the epithelialization process, preventing adhe-

sion to intact tissue and favoring interdigital space care. Non-adhesive hydrocolloid prevents blisters through meticulous skin protection and prevents infections through wound care.<sup>(14,21,30)</sup>

Health education is an important method in nursing care, focusing on psychosocial needs, given that interventions are directed to families to encourage daily activities and leisure activities for children, in addition to seeking rights, as epidermolysis bullosa is also a social phenomenon, which brings with it discrimination. The birth of a child with epidermolysis bullosa can be a traumatic moment for the family. The role of nursing in the care of newborns with epidermolysis bullosa and the family is fundamental and must be started early.<sup>(14,18,19,22,28)</sup>

Nurses are responsible for meeting health education needs, therefore, instructing families, caregivers and/or adolescents regarding topical treatment; handling the baby and child gently so as not to cause new bubbles to appear; care for bathing, feeding, changing diapers, breastfeeding; and vaccination schedule maintenance.<sup>(14,18,19,21,22,28)</sup>

Researchers point out that the recognition of parents regarding the limitations of the disease and in daily life is important for overcoming challenges related to care as well as the social integration of patients and families.<sup>(14,28,29)</sup> Thus, health education carried out by nurses as a means of care on how to deal with children and adolescents with epidermolysis bullosa is paramount.

Therefore, the encouragement given by these professionals to families through communication and qualified listening encourages parents to overcome challenges and reduce psychological trauma, favors coping with the disease of children, adolescents and caregivers, reducing the difficulty of the couple in relation to the feeling of guilt.<sup>(18,19-22)</sup> It is relevant to point out that six articles<sup>(14,17,18-19,21,22)</sup> affirmed the need for nurses to be trained to deal with parents' cognitive and instructional immaturity.

It is worth highlighting the lack of studies regarding the psycho-spiritual care of children and adolescents and their families. It is possible to understand this closer relationship with psychobiolog-

ical needs, since skin impairment triggers numerous systemic changes that can mean a child's life, depending on their age.

Considering the number of results and methodology identified, this research is limited by the absence of experimental studies and randomized clinical trials.

It is important to highlight that only one study<sup>(14)</sup> described the Nursing Process as a guide for assistance to children and adolescents with epidermolysis bullosa and their families. This result is alarming, since, for comprehensive care, it is essential that nurses list Nursing Diagnoses and the other stages of the process to the study clientele. Still, it is known that the Nursing Process is part of assistance in several countries.

It reinforces the need for more research to provide accessible results for care practice, contributing to family members and caregivers and to the care and quality of life of this audience.

## Conclusion

The investigation allowed analyzing NI for the care provided to children and adolescents with epidermolysis bullosa at the hospital level. Surveys report difficulties regarding the availability of materials, treatment and specialized professionals, in addition to limitations in clinical practice related to the characteristics of epidermolysis bullosa. The following nursing care is highlighted: change of decubitus; clothing care; injury site decontamination with hypochlorite solution; sterile covering with essential fatty acids, hydrogel, silicone foam; and analgesia for bathing and dressing change. Avoiding using adhesive material directly on the skin and/or causing shear and changing dressings daily, or as needed or type of foam used are also recommended, in addition to health education strategies aimed at the specificities of epidermolysis bullosa, the rights of this population and psychosocial support for family members with effective interaction. The present study showed the main actions/NI. However, it identified gaps in clinical practice in the care of children and adolescents

with epidermolysis bullosa. Finally, it presents material that should support the treatment and favor nursing care in meeting the basic human needs of this child population. However, there is an urgent need for specific studies that present more specific evidence regarding using dressings in clinical practice with children and adolescents with epidermolysis bullosa, in addition to longitudinal studies on care practices with these families.

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