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Nursing care for a newborn with Lamellar Ichthyosis: a case study in a neonatal unit

Assistência de enfermagem ao recém-nascido com Ictiose Lamelar: um estudo de caso em unidade neonatal

Asistencia enfermera al recién nacido con Ictiosis Lamelar: un estudio de caso en unidad neonatal

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ABSTRACT

Objective: To present the nursing care of a newborn with Lamellar Ichthyosis admitted to the neonatal intensive care unit of a public children's hospital in a municipality in the state of Paraná, Brazil. **Method:** A qualitative intralocal study implementing the Case Study methodology, in which a real case was explored in a delimited system with data collection from multiple sources of information during 66 days of hospitalization in 2016. **Results:** The nursing care present in the protocol was based on: maintaining skin integrity through hydration and continuous lubrication with emollients, temperature control, nutrition and prevention of secondary infections. **Conclusion:** Through the case study, it is affirmed that implementation of the Nursing Process, especially the care plan, was essential for the multidisciplinary success of the treatment. There was improvement in the skin and mucous membranes, as well as prevention of infections, culminating in favorable survival conditions and the autonomy of parents for home care.

DESCRIPTORS

Ichthyosis, Lamellar; Congenital Abnormalities; Infant, Newborn; Intensive Care Units, Neonatal; Neonatal Nursing.

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INTRODUCTION

Lamellar Ichthyosis is an "autosomal recessive inherited disease characterized by defective keratinization and peeling of the epidermis"[1-2]. It is an autosomal recessive genodermatosis which "involves a mutation in the Transglutaminase 1 (TGM1) gene on chromosome 14"[3]. The most severe form of Ichthyosis has an incidence of 1:300,000 births, which is considered to be very low, and has an equal distribution between the genders[4].

Lamellar Ichthyosis appears on the list of the Virtual Health Library's Health Descriptors with the following synonyms: Non-bullous ichthyosiform erythroderma, followed (or not) by the word Congenital; and Harlequin Fetus[5].

It is frequently related to inbreeding, however skin diseases in parents may constitute as possible risk factors for Ichthyosis[6]. The damage is due to the "defect in the transport of intracellular lipids, culminating in the formation of abnormal lamellar granules, which are secreted in the epidermis and determine the appearance of spiny hyperkeratotic scales"[1].

Newborns with Lamellar Ichthyosis have general fine scales on the body, which may be white or gray in color. They may also present ectropion, eclabium, rudimentary ears, alopecia and "problems in plantar and palmar development and movement"[7], due to the lesions that are formed by the skin peeling.

Therefore, Ichthyosis is a dermatosis diagnosed at birth, with treatment having the main goal to guarantee quality of life for the patients. In this process, the nursing team, especially the nurse, stands out among the multiprofessional team for playing a primary role in maintaining skin integrity, preventing infections in hospital and extrahospital care, as well as providing family and caregivers with guidelines and support[8].

Thus, this case study is relevant for addressing a low incidence pathology and disseminating the care quality planned and implemented by a multiprofessional team, focusing on the specific nursing care plan for a newborn with Lamellar Ichthyosis. Furthermore, presenting the development and success of newborn care may impact the support and development of care for others with similar diagnoses and conditions in a Brazilian or international scenario. Thus, the objective of the case study is to present the nursing care of a newborn with Lamellar Ichthyosis admitted to the neonatal intensive care unit of a public children's hospital in a municipality in the state of Paraná.

METHOD

Study design

This is a qualitative study adopting the Case Study methodology[9]. In doing so, the investigator explores a delimited system, focusing on a contemporary event in real life – In this case, the healthcare case of a newborn with Lamellar Ichthyosis. And, as a case study, it covers planning (method that aims at comprehensiveness and effectiveness of integral and multiprofessional care), implementation (through data collection of clinical signs and symptoms) and evolution (with a systematic analysis of the newborn's conditions) of the care plan, taking into account multiprofessional decisions[9-10].

Scenario

The scenario of this case study was a children's hospital located in a municipality in the metropolitan region of Curitiba, Paraná state, southern Brazil. This institution is exclusively public, linked to the Secretariat of Health of Paraná, and is able to serve children from 0 to 18 years old.

At the time of data collection, the institution had the following hospitalization units: 22 beds for surgical pediatrics; 34 for clinical pediatrics; in addition to the beds in the Intensive Care Units: 20 beds in the Neonatal and 10 in Pediatric. There was also a Surgical Center with five active rooms and an Ambulatory service of Medical Specializations, in which Expanded Clinic is practiced. The Physiotherapy Service has a differentiated space structured for child care, receiving referrals from children from the Unified Health System (*SUS – Sistema Único de Saúde*), in addition to those that are being treated in the hospital.

There is a state-of-the-art radiology and imaging service that is a reference center for the care of hospitalized children, outpatients and external appointment by the Unified Health System. The following members of the multiprofessional team operate: nursing technicians, nurses, neonatologists, pediatricians and pediatric specialists (dermatologist, endocrinologist, otolaryngologist, cardiologist, neurologist, ophthalmologist, pediatric gastroenterologist, rheumatologist, infectologist, geneticist, pulmonologist, hematologist, nephrologist, psychiatrist and specialist in inborn metabolism errors), nutritionists, physiotherapists, social workers and speech therapists.

Selection criteria

A newborn (NB) with medical diagnosis of Congenital Lamellar Ichthyosis is constituted in the delimited system[9] or contemporary phenomenon[10], within its real-life context, which means that it is a case according to the Case Study methodology[9]. The NB was hospitalized at the Neonatal Intensive Care Unit (Neonatal ICU) in 2016.

Data collection

Data were collected indirectly from the patient's electronic medical record, the newborn's birth certificate, the Mother's Pregnancy History, as well as from the results of the performed exams.

Data were collected directly through daily clinical evaluation, meaning the complete physical examinations by both the nurse and other professionals of the team, and from the direct care given to the NB. A process of multiprofessional discussion of the case and its evolution was maintained, as well as monitoring with records of all signs and symptoms during the hospitalization period from June to August 2016 (66 days), after being referred from their hometown on the fourth day of life. Some information was also collected directly from the mother. The organization and data collection period for the case study was from January to April 2017.

Data analysis and processing

The data analysis is descriptive, while also using illustrative photos of the evolution of the NB's skin conditions.

Ethical aspects

This study integrates a thematic research project developed at the hospital institution, approved by two Human Research Ethics Committees; one from the Universidade Federal do Paraná, Health Sciences Sector, and the other from the State Health Department. The latter, on August 25, 2016, under Opinion no. 1.698.784, with the consent of the Clinical Board and the Center for Teaching and Research of the Hospital. It followed the criteria of Resolution no. 466 of 2012 of the National Health Council, which deals with research with human beings. As legal guardian for the newborn, the mother was instructed about the research and signed the Informed Consent Form (ICF), allowing the use of images (photographs).

RESULTS

The newborn was born full-term (Parkin 37 + 3 weeks), vaginal delivery, at 12:35pm, weighing 2,160 g with Apgar score 9 and 10, on June 6, 2016, in a city in the interior of Paraná state.

The mother was primigravida, 27 years old, had 10 prenatal consultations, and contained the following information in her Pregnancy Healthbook: negative serology for Human Immunodeficiency Virus (HIV), Syphilis and Hepatitis; Immune toxoplasmosis; obstetric ultrasound without any change. According to the mother's report, the father of the newborn is her first cousin and presented scaly-like skin at birth.

At birth, a collodion membrane was found to envelop the NB. A few hours later, scaly skin lesions were found all over its body. The NB was evaluated by a dermatologist at the Joint Housing of the city where he/she was born and underwent a biopsy on June 7, 2016. Histopathology showed hyperproliferative hyperkeratosis with a thickened granular layer. The dermatologist subsequently prescribed to apply a dressing with moisturizing lotion (Hidrakids/sunflower oil), analgesia with paracetamol drops and eye drops (Lacrifilm) for ocular lubrication. On the third day of life, umbilical catheterization was performed and antibiotic therapy with Ampicillin and Gentamicin.

The newborn was then transferred to the neonatal ICU of the children's hospital of this study on its fourth day of life for follow-up and pediatric dermatological evaluation. Upon admission, the newborn presented "generalized desquamated laminar patches, extensive fissures and hardened membrane (carapace) throughout the body"[1] (Figure 1); "ectropion (turning out of the eyelids) and eclabium (turning out of the lips)"[7].

The guidelines of the pediatric dermatologist consisted of intense skin hydration, without dressing; maintaining body temperature; treatment of secondary infections; care of necrosis of extremities and edema in the atrial region. The ophthalmologic evaluation concluded that there were no corneal lesions, and the prescribed treatment was with Genteal lubricating eye drops every 4/4 h.

After seven days of umbilical catheter use, he/she underwent Peripherally Inserted Central Catheter (PICC) in the cephalic region to maintain intravenous administration of antibiotics. However, due to loss of access, fragile venous network and difficulty in fixation, the patient underwent phlebotomy in the inguinal region E (Figure 2).

The NB was maintained throughout the hospitalization period in a heated and humidified incubator for preventing skin dryness, which was hydrated with liquid vaseline for 58 days. Cetaphil cream (used for hydration and recovery of the damaged skin barrier) was used after this period due to no further peeling.

The NB started treatment with synthetic retinoid on the fourth day of life for keratinization disorder (Acitretin 1mg/kg/day). Treatment was discontinued after 28 consecutive days, as hepatic function alterations were detected. On the 8th day of life (4th day of hospitalization), the NB presented mild respiratory distress and was maintained under oxygen hood therapy for 24 hours.

As the patient initially refused the diet offered orally, an orogastric tube was used (Figure 2). The NB then partially accepted the volume of the oral diet after 16 days of hospitalization. Breast sucking was offered for breastfeeding during the first days of hospitalization, and as he/she presented difficulty in sucking, its mother manually milked her milk to feed it by gavage. However, the mother reported having low milk production after a few days of hospitalization, and so opted for infant formula.

Figure 1 – NB in the neonatal ICU, generalized desquamated laminar patches, children's hospital, Campo Largo, PR, Brazil, 2016.

Figure 2 – Newborn after 10 days of treatment, children's hospital, Campo Largo, PR, Brazil, 2016.

The infant presented a high pain score in the daily nursing records using the Pain Scale adopted in the sector, the Neonatal Infant Pain Scale (NIPS). Fentanyl associated with Dipyrone and Codeine was prescribed during the 60 days of hospitalization.

The patient presented nine infectious processes during the hospitalization period, including bacterial and fungal, and received the following antimicrobials: Ampicillin + Gentamicin; Cefepime + Amikacin; Teicoplanin; Linezolid; Amphotericin; Meropenem + Teicoplanin and Vancomycin.

The multidisciplinary team was attentive in providing humanized care in all processes, with evidence bases which were searched daily. In experiencing their first case of Lamellar Ichthyosis, the nursing assistants were restless to qualify the offered care, to adapt it to the signs and symptoms and to the possible compromises caused by the disease, developing and debating theoretical studies.

The nurses chose the main Nursing Diagnoses through clinical evaluation with daily physical examination to verify new signs for developing the Nursing Process (NP), adopting the Conceptual Model of Wanda de Aguiar Horta as theoretical basis and the Taxonomy I Revised Diagnoses proposed by the North American Nursing Diagnosis Association (NANDA I) in its 2015-2017 classification.

Chart 1 – Nursing Diagnoses and Nursing Prescriptions defined by NICU nurses – Campo Largo, PR, Brazil, 2016.

Nursing Diagnosis	Nursing Prescription
1 – Ineffective infant feeding pattern due to eclabium etiology and skin dryness around the mouth and mucosal lesions.	1 – Administer the gravitational diet. 2 – Maintain elevated decubitus at 30°. 3 – Check the gastric tube positioning before diet administration. 4 – Weigh the newborn daily. 5 – Moisturize lips with Vaseline before administering the oral diet.
2 – Risk of unstable glycemia due to etiology of insufficient gavage diet volume or due to emesis episodes.	6 – Check activity and reactivity of the newborn. 7 – Report capillary glycemia less than 45 mg/dl or greater than 145 mg/dl.
3 – Risk of unbalanced fluid volume due to sepsis etiology and skin lesions.	8 – Monitor skin turgor. 9 – Evaluate mucosal hydration and communicate any changes to the nurse.
4 – Sleep deprivation due to the etiology of irritability and pain.	10 – Keep the environment quiet and dim during the whole period. 11 – Perform restricted and grouped handling.
5 – Ineffective respiratory pattern due the etiology of pain, dyspnea, abnormal respiratory pattern (frequency, rhythm and depth).	12 – Attention to episodes of dyspnea, apnea, nasal fluttering. 13 – Check vital signs every 4 hours.
6 – Risk of paternity or maternity impaired by the etiology of prolonged parental separation and illness.	14 – Encourage the mother and father to care for the newborn. 15 – Maternal/paternal care/holding; kangaroo position. 16 – Permit the parents to stay in the unit indefinitely.
7 – Risk of stress in the caregiver role by the etiology of the illness' severity of the care recipient, health instability of the care recipient.	17 – Maintain active listening. 18 – Ask for psychological support for the parents whenever necessary. 19 – Promote appropriate religious support to the interest and beliefs of the mother and father. 20 – Promote socialization of the companion (mother and/or father) with other companions; encourage short-term exits from the in-hospital environment, taking advantage of open spaces to talk.
8 – Disorganized behavior of the infant due to the etiology in their difficulty to calm down, restlessness and pain. 9 – Acute pain from the etiology of pain evidenced by applying the standardized Checklist of Non-verbal Pain Indicators (CNPI), observation of expressive behavior (shaking, crying) and facial expression of pain.	21 – Promote non-pharmacological measures during painful procedures, such as: rolling over, restraint, non-nutritive suction associated with glucose 25%, skin-to-skin contact for venous punctures, arterial puncture, calcaneal puncture, orogastric tube, upper airway aspiration. 22 – Maintain the newborn properly positioned, nestled in its incubator. 23 – Promote skin-to-skin contact. 24 – Meticulously conduct the pain scale and communicate when a score is equal to or greater than two in order to medicate according to medical prescription.
10 – Injury by impaired primary defense mechanism, such as skin ruptures.	25 – Bath with sterile water and oatmeal daily. 26 – Apply warm compresses with chamomile tea all over the body, every 12 hours. 27 – Moisturize the skin with liquid vaseline every 2 hours.
11 – Risk of infection due to the central venous catheter etiology, desquamated lesions throughout the body extension, oroenteral tube.	28 – Maintain central venous catheter permeability, use aseptic technique to perform flush and medication administration. 29 – Change extensors and stopcocks every 72 hours and diet equipment every 24 hours.
12 – Risk of corneal injury due to ectropion etiology. 13 – Risk of ocular dryness due to vitamin A deficiency.	30 – Sanitize eyes with saline solution. 31 – Apply lubricating eye drops every 4 hours. 32 – Administer Acitretin according to medical prescription.

Thus, after defining the specific Nursing Diagnoses and the corresponding Nursing Prescription, a Care Protocol for Newborns with Lamellar Ichthyosis and their Family was developed and implemented for the newborn and his/her family. This included information about the pathology, aspects of reception and therapeutic relationship with relatives. Moreover, all special care actions for bathing, the skin, mucous membranes and eyes of the newborn were explained.

In summary, the nursing team focused on skin care and comfort of the newborn, developing the following procedures: daily oatmeal and sterile water bath; warm compresses with chamomile tea every 12 hours; care action grouping, because any handling caused irritability; moisturizing and emollient for the skin based on liquid Vaseline every 2 hours, as these promoted "flexibility and movement of the skin to avoid blood circulation restriction"[7]. The professional dermatologist chose Vaseline. In addition, daily skin-to-skin contact with the mother was promoted in order to ease the pain and discomfort of the newborn and to prioritize the newborn bond, involving parents in the care given to the newborn, stimulating skin-to-skin contact for a prolonged period and without schedule restriction, along with valuing and maintaining active listening and psychological support when requested by the parents or when the need was perceived by the team.

The skin care developed by the nursing team was fundamental for improving the "general aspect of skin and lesions"[3], ensured the newborn's survival by preventing complications and promoted skin function as a barrier against infections, as well as favoring healing of the lesions.

The mother was guided and cared for throughout the hospitalization, since the multiprofessional team assumed that parents contribute to the emotional comfort of the newborn during the hospitalization period, in addition to developing care actions that promote them as interlocutors in the care of their child. The parents were accompanied daily by the psychology department so as to provide support to reduce the impact of coping with the health of the child. It was also possible to consult with a geneticist who highlighted the probability of 25% recurrence of the disease in a next pregnancy.

The multidisciplinary team visited the infant/mother on a daily basis to adjust prescriptions and behaviors, and developed periodic discussions on the case, presenting the evolution and the theoretical evidence to support conduct reformulations, as well as diagnoses and prescriptions.

The infant was discharged from the neonatal ICU after 66 days of hospitalization, 70 days old (Figure 3), weighing 2,765 g, and having a height of 51 cm. The outpatient follow-up was scheduled to reinforce "the guidelines, supervision and permanent incentive for continuity of medical treatment and home care"[3].

Figure 3 – Neonatal ICU discharge day, Children's Hospital, Campo Largo, PR, Brazil, 2016.

DISCUSSION

The diagnosis of Lamellar Ichthyosis is predominantly clinical, for which a skin biopsy is considered a complementary examination, and should be performed in the first week of life[11]. The diagnosis in the prenatal period can also be performed by 3-D ultrasonography, capable of identifying signs which are suggestive of Ichthyosis such as eclabium, ectropion, rudimentary ears, contractures and dense floating particles in the amniotic fluid; however, detection of these unusual features requires third-party expertise and are not detectable until the second trimester[11]. Yet, 3-D ultrasonography is not routinely offered by the Unified Health System, and the pathology was not identified in the ultrasound performed by the pregnant woman, but in the biopsy performed on the infant's third day of life.

The neonate presents characteristic facial anomalies of Lamellar Ichthyosis at birth. Ectropion is due to thick corneal extract on the eyelids, responsible for leaving the cornea exposed; a fact which leads to abrasion due to friction, conjunctival edema and corneal ulceration, justifying the need for lubricating eye drops to avoid this complication[12]. Surgical correction of the ectropion has been reported by autografts of the baby's skin and modified human skin[13]. There is no evidence that early surgery results in reducing ectropion from 6 to 12 months of age[14].

When skin integrity is impaired, its function as protective barrier of the organism is affected, and the risk of morbidity and mortality is high. In the case of Ichthyosis, this cutaneous function is markedly impaired, with associated hypernatremic dehydration, impaired thermoregulation, increased metabolic demands, increased corneal extract accumulation and the risk of respiratory failure and sepsis[6,15]. Also, the appearance of cracks as a result of the disease more severely compromises the barrier, which can trigger greater "absorption of physical agents, chemicals and penetration of microorganisms"[3,16] and culminate in septicemia.

The nursing team must act intensely to reduce the chances of infection during hospitalization, thus the main care is aimed at maintaining skin integrity. In this sense, the "maintenance of a preventive insulation environment"[7] which was heated and humidified was prioritized. Therefore, the newborn was kept in a heated and humidified incubator, and vaseline was used on the skin. It is understood that "barrier cream has a lipophilic action, and promotes continuous hydration of intact or injured skin"[3,17], and "body hygiene was performed by means of a daily immersion bath with sterile water plus oatmeal"[3,17]. It is known that oatmeal normalizes the pH of the skin, acts as a sedative and helps relieve irritation and itching[18].

A deep fissure in the epidermis can trigger a chronic pain process, so measures for identifying and measuring pain are indispensable, as are pharmacological measures. Managing the newborn's pain will lead to greater security in the parents. Behavioral scales for evaluating neonatal pain are most effective when they include facial expression,

heart and respiratory rate, crying and agitation. Adequate pain control may require the use of opioids, and consequently there will be a need for some ventilation modality[19]. The severity of pain is attenuated after the superficial layer has been eliminated, and the underlying skin is epithelialized[19].

The use of retinoids in severe cases of Ichthyosis, such as Acitretin derived from vitamin A, acts in the control of epidermal proliferation and differentiation[20-21]. The first successful neonatal use of Acitretin in treating Ichthyosis was reported in 2001, at the dose of 1 mg/kg/day, started on the 10th day of life[21]. Thus, the use of systemic retinoids for managing hospital infection became standard after a reported survival of 83% among 25 treated children, compared to a 24% survival rate of 21 children who did not receive oral retinoid[22]. However, these results should be interpreted with criteria, since half of the untreated babies died within 3 days of birth, i.e. before retinoid therapy[21]. Another study evidenced a 92% survival rate among 12 infants treated with retinoids, when compared to a 50% rate among those untreated[23].

Among the side effects of therapy with oral retinoids are "their toxicity on the skin and mucous membranes"[24], as well as dryness of the lips, mucous membranes and skin are quite frequent. Furthermore, there is "an increase in the serum level of triglycerides and transaminases during treatment, thus monitoring triglycerides and liver function tests of the patients are necessary throughout the treatment"[24].

Electrolytic disturbance is also a complication of transepidermal water loss, which causes severe secondary dehydration and can cause respiratory symptoms[25]. Thus, "signs of skin and systemic infection should be monitored in order to establish the appropriate treatment"[7,25].

There is limited evidence on the benefits of antimicrobial prophylaxis in newborns with Ichthyosis. Authors recommend daily serial surveillance swabs for bacterial and fungal cultures of selected sites (skin folds, nostrils, auditory channels, perianal area) during the first week of life and once per week for the remaining period of stay in the Neonatal ICU[26].

The challenge of early recognition of infection in infants with this pathology is significant, since many findings usually associated with infection in infants (tachypnea, tachycardia, low ingestion) are less specific in Ichthyosis[26].

Inadequate oral intake may be indicative of restricted jaw movement and fatigue in breastfeeding. Whenever there is a high suspicion of infectious disease and a low threshold of laboratory test results (blood and urine), systemic intravenous therapy in neonates with Ichthyosis is promptly initiated[26]. In addition to access for antibacterial treatment, for parenteral nutrition (which was not the case for this baby) and hydration, umbilical catheter insertion is indicated soon after birth. The alternative venous access may be on the scalp, peripheral or through PICC[27]; the latter was adopted to maintain access in the newborn under study.

Most often, newborns with Ichthyosis require supplementary feeding with an oropharyngeal or nasogastric tube. The neonate in the present study received maternal milk from the mother by gavage, since constriction of the eclabium and mandibular mobility made oral feeding difficult. Fluid balance, serum proteins, albumin and electrolytes should be carefully monitored. When adequate sucking and swallowing is established, breastfeeding should be encouraged with the aim of stimulating the bond between mother and child[27]. Formula administration was initiated due to the reduced milk production reported by the mother; a fact that demonstrated fragility in the performance of specialized professionals to stimulate breast milk production, and later breastfeeding.

The absence of thoracic expandability caused by lack of skin elasticity[28] may favor respiratory failure in the newborn with the present condition. In these infants, intubation and mechanical ventilation should be performed in a judicious manner[29].

The prognosis of patients with Lamellar Ichthyosis has considerably improved in recent years, but sepsis is still an important cause of death in the neonatal period[30]. In this sense, it is worth emphasizing the importance of the multiprofessional approach in the neonatal unit, so that there is a favorable evolution and minimization of complications. This perspective contributes to "prolong survival and improve the quality of life of the newborn"[1], as well as the family's view on the pathology.

CONCLUSION

This case study presented the planning, implementation and evolution of the care by a multiprofessional health team, and in particular that of nursing, for a newborn with Lamellar Ichthyosis and their family/mother, admitted to the neonatal intensive care unit of a public hospital in a municipality in the state of Paraná, Brazil. It could be concluded that an integrated team focused on the family and child, as well as on the qualification of care through the best evidence can attain success, even in situations of rare diseases.

The limitation of this study is evidenced by the scarce scientific publication on nursing care of newborns with Lamellar Ichthyosis, thus reiterating that this experience can be replicated in other Neonatal ICU scenarios or for any other rare event.

In view of the incurability of Lamellar Ichthyosis, the main goal of nursing should be to develop preventive actions regarding infectious and ocular symptomatology, concomitantly with the thermal control care of the newborn which focus on the skin/barrier so that it is maintained as stable. Above all, there should be special care with the skin by maintaining use of a topical treatment (moisture control). Application of emollients, moisturizers, lubricants and keratolytics as recommended by the dermatological team, are aimed at maintaining tissue and motor flexibility and reducing pruritus and infectious processes.

The Care Protocol for Newborns with Lamellar Ichthyosis and their family had an impact on the basis and ordering of nursing care and was essential for conducting

multiprofessional treatment of the newborn. With the significant improvement in the skin and mucosal conditions, the prevention of complications and a projection over the possibility of survival were achieved. The family therapeutic relationship is pointed out as an essential factor, with specified guidelines which contributed to treatment adherence and promoting the autonomy of the parents/family to perform home care.

RESUMO

Objetivo: Apresentar os cuidados de enfermagem de um recém-nascido com Ictiose Lamelar internado na unidade de terapia intensiva neonatal de hospital público infantil, em município do estado do Paraná, Brasil. **Método:** Pesquisa de abordagem qualitativa, metodologia de Estudo de Caso, na qual foi explorado um caso real em sistema delimitado, um estudo intralocal, com coleta de dados de múltiplas fontes de informações, em 66 dias de internação, em 2016. **Resultados:** Os cuidados de enfermagem presentes no protocolo basearam-se em: manter a integridade da pele por meio de hidratação e lubrificação contínua com emolientes, controle da temperatura, nutrição e prevenção de infecções secundárias. **Conclusão:** Mediante o estudo do caso, afirma-se que a implementação do Processo de Enfermagem, sobretudo do plano de cuidados, foi essencial para o sucesso multidisciplinar do tratamento. Houve melhora da pele e mucosas, prevenção de infecções, culminando com condições favoráveis de sobrevida e autonomia dos pais para os cuidados em domicílio.

DESCRITORES

Ictiose Lamelar; Anormalidades Congênitas; Recém-Nascido; Unidades de Terapia Intensiva Neonatal; Enfermagem Neonatal.

RESUMEN

Objetivo: Presentar los cuidados de enfermería de un recién nacido con Ictiosis Lamelar hospitalizado en la unidad de cuidados intensivos neonatal de un hospital público infantil, en municipio del Estado de Paraná, Brasil. **Método:** Investigación de abordaje cualitativo, metodología de Estudio de Caso, en la que se exploró un caso real en sistema delimitado, un estudio intralocal, con recolección de datos de múltiples fuentes de informaciones, en 66 días de estancia hospitalaria, en 2016. **Resultados:** Los cuidados enfermeros presentes en el protocolo se basaron en: mantener la entereza de la piel mediante humectación y lubricación continua con emolientes, control de la temperatura, nutrición y prevención de infecciones secundarias. **Conclusión:** Mediante el estudio de caso, se afirma que la implementación del Proceso de Enfermería, sobre todo del plan de cuidados, fue esencial para el éxito multidisciplinario del tratamiento. Hubo mejora de la piel y mucosas, prevención de infecciones, culminando con condiciones favorables de supervivencia y autonomía de los padres para los cuidados en domicilio.

DESCRIPTORES

Ictiosis Lamelar; Anomalías Congénitas; Recién Nacido; Unidades de Cuidados Intensivos Neonatal; Enfermería Neonatal.

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