SYSTEMATIZATION OF CHILD NURSING CARE WITH PYCNODYSOSTOSIS: A CLINICAL CASE REPORT

RESUMO

Objetivo: relatar o caso de uma criança acometida por Picnodisostose, por meio da sistematização da assistência de enfermagem, utilizando a Classificação Internacional para a Prática de Enfermagem (CIPE®).

Método: estudo descritivo e observacional, do tipo caso clínico, desenvolvido em um hospital público no município de João Pessoa/PB, a partir de um instrumento contendo a anamnese, o exame físico e dados registrados no prontuário, utilizando a CIPE® versão 2.0 para determinar os diagnósticos, resultados e intervenções de enfermagem. O estudo foi aprovado pelo Comitê de Ética em Pesquisa, Protocolo nº 222/09.

Resultados: foram identificados cinco diagnósticos de enfermagem prioritários: Risco de trauma; Caminhamento comprometido; Dor musculoesquelética atual; Comportamento Interativo baixo; Ingestão de alimentos comprometida.

Conclusão: a sistematização da assistência de enfermagem possibilitou organização da prestação dos cuidados para uma criança acometida por Picnodisostose, de forma holística e humanizada, levando em consideração a relação criança-família frente à hospitalização.

Descritores: Procesos de Enfermagem; Assistência Integral à Saúde da Criança; Cuidados de Enfermagem; Picnodisostose.

REFERENCES

Albuquerque TM, Oliveira PS de, Silva KL et al. Systematization of child nursing care with Pycnodysostosis through the systematization of nursing care, using the International Classification for Nursing Practice (ICNP®). Method: a descriptive and observational study, of clinical case type, developed in a public hospital in the city of João Pessoa/Paraíba, from an instrument contemplating anamnesis, physical examination and data recorded in the medical record, using ICNP® Version 2.0 to determine the diagnoses, results and nursing interventions. The study was approved by the Research Ethics Committee, Protocol n. 222/09. Results: there were identified five priority nursing diagnoses: risk of trauma; walk compromised; current musculoskeletal pain; bass interactive behavior; food intake compromised. Conclusion: the systematization of nursing assistance enabled the organization to provide care for a child affected by Pycnodysostosis, in a holistic and humane way, taking into account the child-family relationship before hospitalization. Descriptors: Nursing Process; Integral Assistance to Child Health; Nursing Care; Pycnodysostosis.

ABSTRACT

Objective: reporting a case of a child affected by Pycnodysostosis through the systematization of nursing care, using the International Classification for Nursing Practice (ICNP®). Method: a descriptive and observational study, of clinical case type, developed in a public hospital in the city of João Pessoa/Paraíba, from an instrument contemplating anamnesis, physical examination and data recorded in the medical record, using ICNP® Version 2.0 to determine the diagnoses, results and nursing interventions. The study was approved by the Research Ethics Committee, Protocol n. 222/09. Results: there were identified five priority nursing diagnoses: risk of trauma; walk compromised; current musculoskeletal pain; bass interactive behavior; food intake compromised. Conclusion: the systematization of nursing assistance enabled the organization to provide care for a child affected by Pycnodysostosis, in a holistic and humane way, taking into account the child-family relationship before hospitalization. Descriptors: Nursing Process; Integral Assistance to Child Health; Nursing Care; Pycnodysostosis.

SYSTEMATIZAÇÃO DA ASSISTÊNCIA DE ENFERMAGEM À CRIANÇA COM PICNODISOSTOSE: RELATO DE CASO CLÍNICO

SISTEMATIZACIÓN DE LA ASISTENCIA DE ENFERMERÍA AL NIÑO CON PICNODISOSTOSE: REPORTE DE CASO CLÍNICO

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ABSTRACT

Objective: reporting a case of a child affected by Pycnodysostosis, through the systematization of nursing care, using the International Classification for Nursing Practice (ICNP®). Method: a descriptive and observational study, of clinical case type, developed in a public hospital in the city of João Pessoa/Paraíba, from an instrument contemplating anamnesis, physical examination and data recorded in the medical record, using ICNP® Version 2.0 to determine the diagnoses, results and nursing interventions. The study was approved by the Research Ethics Committee, Protocol n. 222/09. Results: there were identified five priority nursing diagnoses: risk of trauma; walk compromised; current musculoskeletal pain; bass interactive behavior; food intake compromised. Conclusion: the systematization of nursing assistance enabled the organization to provide care for a child affected by Pycnodysostosis, in a holistic and humane way, taking into account the child-family relationship before hospitalization. Descriptors: Nursing Process; Integral Assistance to Child Health; Nursing Care; Pycnodysostosis.

RESUMEN

Objetivo: presentar un caso de un niño afectado por Picnodisostosito a través de la sistematización de la asistencia de enfermería, utilizando la Clasificación Internacional para la Práctica de Enfermería (CIPE®).

Método: un estudio descritivo y observacional del tipo de caso clínico, desarrollado en un hospital público en la ciudad de João Pessoa/Paráiba, a partir de un instrumento que comprende la anamnesis, el registro físico y los datos registrados en el prontuario, utilizando la CIPE® versión 2.0 para determinar los diagnósticos, resultados e intervenciones de enfermería. El estudio fue aprobado por el Comité de Ética en Investigación, Protocolo nº 222/09. Resultados: se identificaron cinco diagnósticos de enfermería prioritarios: Riesgo de trauma; Caminar comprometido; Dolor musculoesquelético actual; Comportamiento Interactivo bajo; Ingestión de alimentos comprometida. Conclusión: la sistematización de la asistencia de enfermería permitió la organización de la prestación de los cuidados para una niño afectado por Picnodisostosito, de forma holística y humanizada, teniendo en cuenta la relación niño con la familia frente a la hospitalización. Descritores: Procesos de Enfermería; Asistencia Integral a la Salud de la Crianza; Cuidados de Enfermería; Picnodisostosito.

REFERENCES

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INTRODUCTION

The change in the number of morbidity and mortality in the general population, highlights the increasing cases of chronic degenerative diseases, necessitating the demand for services and more complex actions, with rising costs, need for new drugs and new technologies. 1

The advancement of genetics in health practice has become significant in relation to the understanding, diagnosis and treatment of pediatric diseases in the past few decades. Many of these little-known before now can be understood, as is the case of Lysosomal Diseases (DL), which are pediatric neurodegenerative diseases. 2

The understanding of the pathophysiology of most of these conditions allowed the establishment of protocols that minimize or even avoid these clinical manifestations.3

Among the DL of larger approach highlights the Pycnodysostosis, also known as mucopolysaccharidosis (MPS), which is a rare hereditary disease, autosomal recessive with the exception of type II (also called Hunter Syndrome). The MPS is defined as a genetic alteration related to the X chromosome and can be classified as mild or severe, according to the involvement of the central nervous system, caused by specific lysosomal enzyme deficiency that acts sequentially in the degradation of glycosaminoglycans (GAG) components of the membrane and extracellular matrix. The enzyme deficiency in varying degrees triggers the accumulation of glycosaminoglycans in the lysosome causing cellular dysfunction in multiple organs.4-5

The diagnosis is usually performed by a clinical geneticist through specific tests that verify the pattern of GAGs in the urine, confirmed by analysis of lysosomal enzymes in blood leukocytes or skin fibroblasts and genetic study. In cases of suspected during pregnancy, amniocentesis and chorionic villus biopsy allows the diagnosis during prenatal.6

The clinical presentation can be varied, the most common manifestations: skeletal changes, with consequent bone fragility fractures, heart problems, mental retardation, organomegaly and facies characteristics. The changes of these patients otorhinolaryngologic can be divided into three groups: ear disorder, adenotonsillar hypertrophy, changes in eating and chewing difficult airway changes. Moreover, recurrent infections of the upper respiratory tract may also be causes of tonsil disease associated framework.6

In order to achieve the holistic, humane and effective care, paying attention to the specifics of a chronic degenerative disease, it is pertinent to stress the use of the Nursing Care System (NCS), which can be understood as a methodology to organize and systematizing care based on scientific knowledge. Aims to identify situations of the health-disease process and the needs for nursing care, but also to subsidize intervention involving the promotion, prevention, recovery and rehabilitation of the health of the individual, family and community.7

Resolution No. 358 of the Federal Council of Nursing (COFEN) 2009 recommends that the NCS should be performed in all health institutions in the professional nursing care occurs. It is operationalized through the nursing process consists of five steps: history of nursing, nursing diagnosis, nursing planning, implementation and evaluation of nursing.8

The International Classification for Nursing Practice (ICNP®) is used as a unified language that contemplates the phenomena, nursing interventions and outcomes as primary elements of its construction. Such terminology is standardized internationally and was developed by the International Council of Nurses (ICN) in 1989, together with researchers from classification systems in nursing recognized by the American Nurses Association (ANA). Its composition provides playback of a common language to best describe the practice of nursing in the health information systems.9

Then you realize the significance of a special care to children affected by this disease, making it necessary to systematize nursing care, using the International Classification for Nursing Practice (ICNP®) in order to develop nursing diagnoses, the order to offer a humanized, individualized and systematic care.9

OBJECTIVE

● Reporting the case of a child affected by Pycnodysostosis through the systematization of nursing care, using the International Classification for Nursing Practice (ICNP®).

METHOD

Descriptive, observational study of the clinical case of an eight-year-old female diagnosed with Pycnodysostosis, hospitalized in the pediatric clinic of a public hospital in the city of João Pessoa type-PB, in May, 2011.

Data were collected from the care provided to children and their families, with the
development of the diagnostic procedures for performing the general and specific physical examination, aided by a structured questionnaire, covering basic human needs described by Horta, and other recorded data in the child's records and interviews with patients and their families.

From the information collected it was possible to guide the choice of the most appropriate nursing diagnoses and care, with the use of ICNP® (version 2.0).

To assist the development of such diagnostic strategy was used as the conceptual map, which corresponds to a tool that allows enhancing the content fetched by the researcher ensuring continuous progression in knowledge acquisition, capable of providing identification data of interest and their inter-relationships for developing core diagnostic. For the elaboration of the concept map was used CMap Tools software in its version 5.03, developed by the Institute for Human Machine Cognition, University of West Florida, which allows the user to build knowledge models represented by conceptual maps of their patent. With the definition of nursing diagnoses, proceeded to care planning and therefore the implementation of child care.

It is noteworthy that the data collection was initiated only after clarification of the research and signed the consent form and the mother Free and Informed Consent of the child, in accordance with Resolution nº 466/12 of the National Health Council.

This study is linked to a research project developed with children and adolescents with chronic illness, approved by the hospital's Research Ethics Committee under protocol nº 222/09, which is associated with the Group of Studies and Research in Child and Adolescents and the Group of Studies and Research Grounds of Nursing Care, the graduate program in Nursing, Federal University of Paraíba/PPGENF/UFPB.

RESULTS

◆ Case Study - Historic

Student, Y.S.C, eight, female, Catholic, daughter of parents with first-degree kin cousins, born in Banana-PB, residing in rural areas of this county, admitted to the pediatric clinic of a teaching hospital in João Pessoa-PB, accompanied by their mothers’, in April 2011, where he received medical diagnosis for Pycnodysostosis. Born preterm, with 1,500g, due to loss of amniotic fluid, by cesarean section, not cried at birth, presenting cyanotic.

The initial historic of hospitalization: According to mothers’, the school began to show high bone fragility, with a history of several fractures in different parts of the body - not knowing specify the age at which the child was when the first fracture appeared, six months before the hospitalization, fractured femur which caused difficulties in walking. Because of this weakness , body aches to feel jerks movements, getting underactive and preventing people to approach her for fear of “breaking their bones.” The mother, to ease the pain process, provides, by itself, has anti-inflammatory, as well as the prohibition of performing games that require movement. It is emphasized that the school has open cranial sutures and fontanelles with bregmatic lambdoid palpable, but also failure to thrive. Moreover, it has difficulty in chewing due to the irregular layout of your teeth.

Before the picture presented by the school and by not getting medical diagnosis defined in the health services who attended for several years, the child’s mother decided to seek the hospital school, since this is a reference in care for children with chronic illness, with indefinite diagnosis.

The examination and evaluation of the needs of the school: Vital signs and anthropometric data: T=36,8°C, HR : 96 bpm RR: 22 bpm; Height 1.04 m, Weight: 17 kg; Fontanel bregmatic: 8x5 ppd; Fontanel lambdoid: 4x3 ppd; Need for Neurological Regulation: conscious, oriented, non-communicative, underactive; Need for sleep and Rest : sleep and rest preserved nutritional Need: decreased food intake at breakfast, did not accept the diet (SIC mother); Need for Body Lotion: body care satisfactory; Need for Physical Integrity and Cutis : Skin and normocollorated and hydrated, acyanotic, anicteric mucous, afebrile, turgor and preserved elasticity; Need oxygenation: chest up, bilateral chest expansion, eupneic, MV+, rhythmic breathing; Need Vascular regulation: Normocardic, RCR in Q2, BCNF in all outbreaks of auscultation; Need for Elimination: flaccid abdomen , RHA decreased in all quadrants, not referred pain to the superficial and deep palpation, presence of hollow sounds in all quadrants, eliminations bowel and bladder present with normal characteristics (SIC mother); Need bw/mobility/motility mechanics: difficulty in walking, with a limp because of the recent fracture of the femur ; Need for pain perception, which refers to pain in the body jerks, getting, thus underactive; Need for Communication/Recreation and Leisure: Low social interaction; Need
Love/Gregarious: Presented good relationship with the mother and reported feeling homesick father and brothers. The needs for shelter and housing, housing is brick, with four rooms (one room, a bedroom, kitchen and bathroom), with ten residents, clean water, electricity, sewage, garbage collection, with pets, puppy and cat.

Based on clinical history presented, we sought to develop a conceptual map to explain the decision making regarding the priority nursing diagnoses, as shown in Figure 1.

Figure 1. Diagrammatic representation of data concerning the clinical case that served as a basis for identification of nursing diagnoses according to the ICNP ®. João Pessoa, PB, 2011.

After the identification of nursing diagnostic statements, was traced planning of nursing care, as shown in Figure 2.

<table>
<thead>
<tr>
<th>Nursing diagnoses/Expected results</th>
<th>Nursing interventions</th>
<th>Evolution</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Risk of trauma/Avoid the emergence of trauma</strong></td>
<td>Guide the school through playful, to replace activities that require sudden movements and direct contact (e.g. racing, playing on playgrounds and leisure) for those that require gentle movements (painting, drawing). Guide to escort to keep the bars of the bed raised, when the child is sleeping; place the auxiliary ladder when need to rise or fall of the bed; keep the environment free of objects and materials that can cause falls.</td>
<td>During the study, the child did not present any kind of trauma.</td>
</tr>
<tr>
<td><strong>Walk compromised/Improve the walk</strong></td>
<td>Encourage frequent visits to the recreation sector, thus enabling progressive, hiking. Demonstrate through drawings, illustrations and/or videos, secure forms of wandering, without risk of falls. Guide the escort about the importance to be always next to the school during ambulation, offering support, safety and comfort. Recommend to the Physiotherapy service.</td>
<td>After a day of intervention, with the help of escort and after session of physiotherapy, the child spent wandering three times a day, number, and compatible with the visits to the recreation sector.</td>
</tr>
<tr>
<td><strong>Current Musculoskeletal pain/Submit absence or reduction of musculoskeletal pain</strong></td>
<td>Guide the child through toys, to prevent sudden movements, replacing them with smooth movements. Apply non-pharmacological techniques for pain relief (e.g. cold compresses, hot, massages).</td>
<td>One day after the intervention, was referred to reduction of musculoskeletal pain.</td>
</tr>
<tr>
<td><strong>Interactive behavior under/Introduce greater interaction with the health team as well, with children recreation</strong></td>
<td>Using the technique of therapeutic toy, before performing procedures. Motivate ludic activities (paint, draw, make collages, cutouts). Encourage participation in activities held in recreation. Encourage participation of escort along with the child in leisure activities. Recommend psychology service.</td>
<td>After a day of intervention, the child appeared more communicative and interactive with the health team and with children to visit the recreation sector.</td>
</tr>
<tr>
<td><strong>Food intake compromised/Introduce improved food intake</strong></td>
<td>Interact with the service of nutrition, requesting the proper nutrition to children's health conditions, as food of Pasty consistency. Recommend to the dental service. Recommend to the speech therapy service.</td>
<td>On the same day of the intervention, the child accepted well the diet.</td>
</tr>
</tbody>
</table>

Figure 2. Planning of nursing care to a child with a diagnosis of Maroteaux-Lamy syndrome, according to the ICNP ® version 2.0. João Pessoa, PB, 2011.
DISCUSSION

The Pycnodysostosis is a universal autosomal recessive disease occurrence and association with high inbreeding, due to their genetic heritage. Belongs to the group of skull-tubular bone dysplasia, in which the bones of those affected become progressively sclerotic, essentially due to a failure in osteoclast activity by mutations in the gene encoding cathepsin K enzyme, located on chromosome 1q21, causing osteosclerosis. Thus, there is increased bone density, resulting from minimal trauma fractures, particularly in the long bones - actually occurred with the school, which had a fracture in the femur - which justifies the nursing diagnosis of Risk for Trauma.15

Fractures are the main impairment in people diagnosed with Pycnodysostosis, being paramount, so care aimed at prevention of trauma. Such precautions include careful handling of an affected child, along with the combination of exercise and insurance activities, which do not require much impact.16

The recent fracture of the femur associated with the picture of pain of the musculoskeletal system, which is characterized by stiffness and joint deformities in people with Pycnodysostosis, converged on the school presented difficulties in walking, which resulted in the nursing diagnoses committed Walking and Musculoskeletal Pain current, respectively.17

The period between seven and ten years old, agreed to be understood as school period is marked by significant changes in the cognitive, social-emotional and communication aspects of plastic, since this stage of the child’s reasoning is more logical, more understanding of the facts. Thus, we believe that the nursing diagnosis of low interactive behavior, besides being associated with clinical conditions of Pycnodysostosis, also has ties to the stage of child development affected by hospitalization experienced by the child.18

Hospitalization is for the child experienced a difficult experience because instead of your daily routine, the environment they are already familiar and the presence of relatives and friends, the child is exposed to routine procedures that cause you discomfort, in an impersonal and stressful environment, surrounded by strangers. This can leave the anxious, insecure and scared child, especially when there is a preparation for hospitalization, as well as for the treatment to be undertaken.19-20

The play allows a child’s learning as well as being critical at all stages of its development, as it allows her to relate to other children, thus returning to “their world”, but also is a way to keep her from suffering.21

The use of therapeutic play proved to be an effective strategy in reducing the traumatic effects of hospitalization, as it helps the child to understand the reality we are experiencing, to assimilate new situations, to know the importance of each procedure performed, to express their feelings pain, fear, anxiety. Thus, this technique of playing encourages social, emotional, intellectual and therapeutic development, from the time that reduces negative feelings about hospitalization.20,22

It is noteworthy that the illness and hospitalization of children change family dynamics, making a favorable family to develop feelings and emotions such as sadness, fear, shame, guilt, helplessness. In this sense, it is understood that the figure of the person accompanying the child during hospitalization, most often represented by the mother, needs attention and care of the health team as much as the child, since the welfare of a directly affects the condition of the other.19,20

Regarding the nursing diagnosis impaired food intake, the literature shows that people diagnosed with Pycnodysostosis may show changes in the dental arch, tongue, chewing and swallowing. These factors may contribute to a reduced food intake, affecting nutritional status, resulting in increased incidence of complications secondary to that underlying disease. Thus, consumption of foods pasty is considered the most appropriate for children who have oral motor dysfunction.5

The nurse is the professional health team that spends more time near the child and family and, consequently, presents comprehensive view of the needs of both. Thus, standardization of individualized care is shown as a needed to underpin the clinical nursing practice strategy as it contributes both to the establishment of nursing interventions and the evaluation itself. As the nurse when performing planning, ensures quality care and effectiveness, therefore, allows you to diagnose the child's needs, ensures proper prescription of care, guidance supervision of staff performance, evaluating the results and the quality of assistance as guides the actions.21-4
CONCLUSION

This study allows us to understand the key needs presented by a child Pycnodysostosis, since it is a rare and scarce national journals that address the theme, especially with regard to nursing disease.

The systematization of nursing care (SAE) through the nursing process, enabled the achievement of positive elements in the participating child care focused study and his family, since after the implementation of the proposed interventions in the planning, it was possible to achieve expected for diagnostic results. This demonstrates that when applied in professional practice, NCS assists nurses to develop a care grounded in scientific knowledge, qualifying and optimizing the work process and thus with the possibility of ensuring a humane, comprehensive and holistic care for the binomial mother-child.

REFERENCES
Importance of using therapeutic toys in care of children with Diabetes type 1.


Systematization of child nursing care with...