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ISOLATED ROBIN SEQUENCE: NURSING DIAGNOSES¹

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ABSTRACT

Objective: to identify nursing diagnoses in infants with isolated Robin Sequence.

Method: a descriptive study developed in a hospital specialized in craniofacial anomalies and related syndromes, from November 2015 to March 2016. Twenty infants with isolated Robin Sequence participated in the study. For the data collection, the nursing history was used. The nursing diagnoses were listed according to Nanda-International. To compose the results, a descriptive statistical analysis was used.

Results: eight diagnoses were identified, five of which focused on the problem and three on risk factors, including: risk of aspiration (n=20, 100%); risk of infection (n=20, 100%); ineffective breastfeeding (n=19, 95%); unbalanced nutrition lower than body needs (n=19, 95%); ineffective infant feeding pattern (n=19, 95%); risk of damage to skin integrity (n=18, 90%); ineffective airway clearance (n=11, 55%) and impaired spontaneous ventilation (n=11, 55%).

Conclusion: infants with isolated Robin Sequence presented nursing diagnoses that were related to respiratory, feeding, safety and comfort problems and favored the planning and implementation of nursing care when tracing a care profile.

DESCRIPTORS: Nursing Diagnosis. Pierre Robin Syndrome. Nursing. Nursing Processes. Pediatric Nursing.

SEQUÊNCIA DE ROBIN ISOLADA: DIAGNÓSTICOS DE ENFERMAGEM

RESUMO

Objetivo: identificar os diagnósticos de enfermagem em lactentes com Sequência de Robin isolada.

Método: estudo descritivo desenvolvido em um hospital especializado em anomalias craniofaciais e síndromes relacionadas, no período de novembro de 2015 a março de 2016. Participaram 20 lactentes com Sequência de Robin isolada. Para a coleta de dados utilizou-se o histórico de enfermagem. Os diagnósticos de enfermagem foram elencados segundo a Nanda-Internacional. Para confecção dos resultados utilizou-se a análise estatística descritiva.

Resultados: foram identificados oito diagnósticos, sendo cinco com foco no problema e três de risco, incluindo: risco de aspiração (n=20, 100%); risco de infecção (n=20, 100%); amamentação ineficaz (n=19, 95%); nutrição desequilibrada menor que as necessidades corporais (n=19, 95%); padrão ineficaz de alimentação do lactente (n=19, 95%); risco de integridade da pele prejudicada (n=18, 90%); desobstrução ineficaz das vias aéreas (n=11, 55%) e ventilação espontânea prejudicada (n=11, 55%).

Conclusão: lactentes com Sequência de Robin isolada apresentaram diagnósticos de enfermagem que se relacionaram a problemas respiratórios, alimentares, de segurança e conforto e favoreceram o planejamento e implementação dos cuidados de enfermagem ao traçarem um perfil assistencial.

DESCRIPTORIOS: Diagnóstico de enfermagem. Síndrome de Pierre Robin. Enfermagem. Processos de enfermagem. Enfermagem pediátrica.

SECUENCIA DE ROBIN AISLADA: DIAGNÓSTICOS DE ENFERMERÍA

RESUMEN

Objetivo: identificar los diagnósticos de enfermería en lactantes con Secuencia de Robin aislada.

Método: estudio descriptivo desarrollado en un hospital especializado en anomalías craneofaciales y síndromes relacionados, en el período de noviembre de 2015 a marzo de 2016. Participaron 20 lactantes con Secuencia de Robin aislada. Para la recolección de datos se utilizó el histórico de enfermería. Los diagnósticos de enfermería fueron enumerados según la Nanda-Internacional. Para la confección de los resultados se utilizó el análisis estadístico descriptivo.

Resultados: fueron identificados ocho diagnósticos, siendo cinco con foco en el problema y tres de riesgo, incluyendo: riesgo de aspiración (n=20, 100%); riesgo de infección (n=20, 100%); lactancia ineficaz (n=19, 95%); nutrición desequilibrada menor que las necesidades corporales (n=19, 95%); el patrón ineficaz de alimentación del lactante (n=19, 95%); riesgo de perjuicio a la integridad de la piel (n=18, 90%); desobstrucción ineficaz de las vías aéreas (n= 1, 55%) y ventilación espontánea perjudicada (n=11, 55%).

Conclusión: lactantes con Secuencia de Robin aislada presentaron diagnósticos de enfermería que se relacionaron con problemas respiratorios, alimentarios, de seguridad y confort y favorecieron la planificación e implementación de los cuidados de enfermería al trazar un perfil asistencial.

DESCRIPTORES: Diagnóstico de enfermería. Síndrome de Pierre Robin. Enfermería. Procesos de enfermería. Enfermería pediátrica.

INTRODUCTION

The Robin Sequence is a malformation characterized by a triad composed of micrognathia (lower jaw abnormally smaller than normal) and glossoptosis (abnormal repositioning of the tongue), most often accompanied by cleft palate. It can be presented in isolation, and called isolated Robin Sequence (IRS), or in association with syndromes and other clinical comorbidities.¹ The etiology is multifactorial and the incidence varies from 1:8500 to 1:14000 live births.²⁻³

The symptoms arising from the triad include respiratory obstruction and eating problems, requiring immediate diagnosis to target treatment in order to avoid complications that include death. The treatment must be holistic and based on a multidisciplinary team that includes nursing, favoring a qualified care to the patient and their family and positively influencing the treatment.⁴⁻⁵

Due to respiratory and eating problems, infants with IRS often require artifacts that include nasopharyngeal intubation and feeding tube.⁶⁻⁷ However, the use of these does not contraindicate the hospital discharge, as long as that their parents or caregivers are trained to maintain the care at home. In this context, nursing plays a prominent role; in which, in addition to providing direct care to infants, it is responsible for training family members for providing care.⁸⁻¹⁰

Several instruments have been presented in nursing to systematize care. Among them, the Nursing Process should be highlighted; it is defined as an important instrument that assists nursing professionals in the planning and implementation of interventions to obtain the expected results. It is based on clinical reasoning and includes the phases of Nursing History or

Research (anamnesis and physical examination), Nursing Diagnosis, Planning, Implementation or Prescription and Evaluation of the results.¹¹

The phases of the Nursing Process are interconnected. In order to formalize the Nursing Diagnoses it is indispensable that the Nursing History does not contain knowledge gaps and/or misinformation. The Nursing Diagnosis consists of a clinical judgment performed exclusively by nurses, concerning a human response to real or potential health/life conditions, which are eligible for an individual, family, group or community, and are the basis for the selection of actions or interventions to achieve the expected results.¹¹⁻¹²

Over time, it was observed the need to develop standardized languages that facilitate the implementation and formalization of the nursing process in different contexts. Thus, among others, the classification of the Nursing Diagnostics of NANDA-International emerged, which has been translated into several languages and has been used worldwide.¹²

In this context it is questioned: what are the eligible nursing diagnoses in infants with isolated Robin Sequence? In the databases consulted, only one study on nursing diagnoses in infants with isolated Robin Sequence was found, in a case study format, in which the authors point out the need for new studies with larger casuistic that favor the generalization of the results.¹³

It is expected from this publication, through the identification of nursing diagnoses, to offer subsidies to plan and implement interventions that facilitate the process of rehabilitation of these infants, in addition to promoting holistic, quality, safe and humanized care. It should be emphasized that decentralization policies proposed by

the Unified Health System focus on the care of these infants in different services and different realities. Thus, the dissemination of scientific knowledge on nursing care will certainly reflect benefits both for patients and families and for health professionals.

Thus, the objective of this investigation was to identify the nursing diagnoses in infants with IRS.

METHOD

A descriptive study developed at a national and international reference hospital in the treatment of craniofacial anomalies and related syndromes, more precisely in the Semi-Intensive Care Unit. This unit is composed of eight beds intended for the care of newborns and infants, who usually present respiratory and feeding problems. In addition, this unit is focused on the training of parents and caregivers in order to maintain the provision of care at home.

Considering the stipulated period of data collection, twenty infants with characteristic signs and symptoms of IRS, that is, micrognathia, glossoptosis with or without cleft palate, were included. Infants with Robin sequence associated with syndromes or other abnormalities, or comorbidities, such as neurological and/or cardiac disorders, were excluded.

After the definition of the eligible cases, the legal guardian of the child was invited to join the survey, and this approach was carried out in a reserved and timely manner. The data collection was performed from November 2015 to March 2016. The data were analyzed by the Excel 2010 Program and submitted to a descriptive statistical analysis.

The research began after the approval of the Research Ethics Committee involving Human Beings from the study scenario, with approval CAAE: 49104315.4.0000.5441. The responsible for the infant formalized the adherence to the research through the signing of the Free and Informed Consent Term.

The data collection was performed independently by two nurses, authors of the present study, who were duly trained to do so, in order to offer greater trustworthiness to the results. For the Nursing History, the anamnesis was performed through the collection of data in the medical record, in addition to interviewing parents or guardians in a reserved environment. Concomitantly, a complete physical examination of the child was performed. For this, a data collection instrument was used by the authors. Once the Nursing History was carried out, the Nursing Diagnostics was formalized according to the Nanda-International Taxonomy.¹²

The diagnoses listed by both researchers were immediately eligible. In case of disagreement among the evaluators, there was a reevaluation of the process by a third nurse, also author of this study, with greater clinical experience, in order to obtain consensus.

RESULTS

Twenty patients with mean age of 22 days (± 8) were assessed, of which 12 (60%) were females. Regarding the socioeconomic classification, the prevalence was low ($n=10$; 50%) followed by the mean ($n=6$; 30%). The mean time of hospitalization was 7.2 (± 1.6) days.

The problem-focused diagnosis, its factors related to their respective defining characteristics are presented in table 1.

Table 1 - Frequency of the nursing diagnoses with focus on the problem, its defining characteristics and related factors. Bauru, SP, Brazil, 2016. (n=20)

Problem-focused diagnosis	n	%
Ineffective breastfeeding (00104)		
Defining characteristics		
Unsupported breast suction	15	75
Absence of infant weight gain	1	5
Related factors		
Oropharyngeal defect	19	95
Unsatisfactory reflection of infant suction	19	95

Problem-focused diagnosis	n	%
Unbalanced nutrition: less than the bodily needs (00002)		
Defining characteristics		
Perception of the inability to ingest food	19	95
Insufficient muscle tone	18	90
Body weight 20% or more below ideal	1	5
Related factors		
Inability to ingest food	19	95
Ineffective infant feeding pattern (00107)		
Defining characteristics		
Incapacity and coordination of sucking, swallowing and breathing	19	95
Inability to maintain an effective suction	19	95
Inability to initiate an effective suction	15	75
Related factors		
Oropharyngeal deformity	19	95
Prematurity	2	10
Ineffective airway clearance (00031)		
Defining characteristics		
Dyspnea	11	55
Change in the respiratory rate	11	55
Respiratory adventitious noise	4	20
Related factors		
Foreign body in the airway	11	55
Excessive mucus	5	25
Impaired Spontaneous Ventilation (00033)		
Defining characteristics		
Dyspnea	11	55
Increased use of accessory muscles	11	55
Increased heart rate	8	40
Related factors		
Respiratory muscle fatigue	11	55

The risk diagnosis with their respective risk factors are presented in table 2.

Table 2 - Frequency of the nursing diagnoses of risk with their respective risk factors. Bauru, SP, Brazil, 2016. (n=20)

Risk Diagnosis	n	%
Risk of aspiration (00039)		
Risk factors		
Enteral feeding	19	95
Presence of oral/nasal tube	18	90
Presence of intra-tracheal tube	10	20

Risk Diagnosis	n	%
Neck surgery	2	10
Risk of infection (00004)		
Risk factors		
Invasive procedure	20	100
Inadequate vaccination	6	30
Malnutrition	1	5
Risk of injury to the skin integrity (00047)		
Risk factors		
Extremes of age	20	100
Mechanical factors	18	90
Inadequate nutrition	1	5

DISCUSSION

When analyzing the characterization of infants in relation to gender, it was observed the predominance of females. A research has indicated the prevalence of 65% of isolated cleft palate in this gender.¹⁴ Among the infants affected with IRS, 90% have cleft palate, justifying this result.²

As for the socioeconomic class, the low class has prevailed. It is inferred that this result is based on the fact that the hospital is public, attending exclusively the patients of the unified health system, therefore, the less favored social class.⁹

The mean age of the infants with IRS found reflected the need for an early hospitalization, considering the complications, among them death. The late hospitalization may cause the infant to have the worsening of the symptoms, such as respiratory failure and malnutrition associated with dysphagia.⁶⁻⁷ The literature indicates that infants with IRS have a good prognosis when treated early.⁴⁻⁷

Children with IRS usually have breathing problems and eating difficulties.⁶⁻⁷ In the casuistic of the present study, the following nursing diagnoses related to breathing were listed: "risk of aspiration", "impaired spontaneous ventilation" and "ineffective airway clearance". A similar result was evidenced in another study.¹³

The nursing diagnosis "Risk of Aspiration" is defined as vulnerability to the entry of gastrointestinal secretions, oropharyngeal secretions, solid or liquid in the tracheobronchial pathways, which may compromise health.¹² Most of the

infants presented this diagnosis. Among the risk factors contemplated by the diagnosis, the enteral feeding prevailed in this study. Similar results were observed in other studies.⁷⁻⁹ In infants with IRS, the oral intake is generally impaired as a result of a respiratory obstruction that hinders the coordination of sucking, swallowing and breathing. The micrognathia and glossoptosis impair the anteriorization of the tongue; which, along with the palate cleft, results in lower intraoral negative pressure, resulting in the need for feeding through the use of a tube, increasing the risk of bronchoaspiration.^{6,15}

It should be highlighted that for the occurrence of pneumonia, the broncho-aspiration of a large volume is not necessary. Micro-aspirations are indicated as an important etiological agent.¹⁶ The maintenance of the elevated decubitus from 30 to 45° reduces the incidence of complications, such as vomiting and regurgitation that can lead to pulmonary broncho-aspiration. This intervention, associated or not to others, reduces costs, improves the survival rate, promotes shorter hospital stays, and minimizes costs.¹⁷⁻¹⁸

The most severe cases of IRS are predisposed to the gastroesophageal reflux disease associated with intra-thoracic negative pressure due to inspiratory effort.⁶⁻⁷ However, they present improvement with the treatment of the respiratory obstruction.¹⁹

Different treatment modalities have been described in the literature to relieve the respiratory obstruction and to improve the feeding difficulties in infants with IRS. The priority is to maintain the

permeability of the upper airways. Among the non-surgical techniques, the positioning in the ventral decubitus is present, being effective for infants with a mild respiratory obstruction, since this is caused by the retro-position of the tongue. This treatment modality facilitates breathing, as it promotes a cervical hyper extension, however, it is not sufficient for moderate or severe cases.⁶⁻⁷

If the result with the positioning is not effective, another conservative method is used: the nasopharyngeal intubation (NI).⁶ It is a simple and minimally invasive method to promote the airway permeability in infants with IRS through the introduction of a 3.0 cm to 3.5 cm diameter silicone orotracheal intubation cannula from the nose to the nasopharynx. A thin wire drawn from the gauze should be placed in front of the outside of the tube to check its permeability.⁶⁻¹⁰ The cannula positioned below the tongue will overcome the mechanical barrier imposed by it, favoring the ventilation.²⁰

The NI improves the respiratory obstruction caused by the posterior tongue traction and is effective until sufficient growth occurs so that respiratory and feeding difficulties are overcome, avoiding surgeries such as the tracheostomy that is indicated for severe cases and the mandibular osteogenic distraction, in well selected cases.^{6,20}

A retrospective study concluded that most of the infants with IRS were treated conservatively, with NI being the most commonly used method. The infants were hospitalized for a short period and there was no death, reflecting the efficiency of the procedure associated with an interdisciplinary treatment.⁷

It is important to highlight that the NI is considered a simple procedure that does not contraindicate the hospital discharge, on the contrary, it is taught to parents or caregivers during the hospitalization period, so that it can be continued at home, reducing the hospitalization time.^{6-7,10} This treatment modality is efficient both for the recovery of respiratory difficulties and for helping to improve eating difficulties.⁶⁻⁷

The nursing diagnosis "Ineffective route clearing" is related to the inability to eliminate secretions or obstructions of the respiratory tract necessary to keep the airway unobstructed.¹² The mandibular hypoplasia and the tongue retro-positioning cause respiratory dysfunctions by reducing the air passage space, thus hindering the respiratory process, requiring the use of NI in order to provide adequate gas exchange.^{1,4,6}

This result is in line with what has been found in another study.¹³

In order to increase the chest expansion and relieve the sensation of dyspnea, the angle of inclination of the head is raised, since the upright position favors the pulmonary expansion, promoting maximal inspiration and improving the alveolar oxygenation.²¹ In order to suppress the deficiency in the diffusion of the gases, the organism increases the pulmonary ventilation. Thus, there is an increase in the respiratory rate and depth, with the activation of the accessory musculature of the breath. After the activation of these mechanisms of compensation, the individual presents a sensation of respiratory discomfort, focusing on dyspnea.²²

The factors that influence a greater occurrence of the diagnosis of ineffective airway clearance are the narrow airway and propensity to secretion retention, since the infant's respiratory system is immature. In the present study, the narrowing is explained by the micrognathia and glossoptosis that reduce the nasopharyngeal space.

The diagnosis of the airway obstruction mechanism is important to direct the infant's modality of treatment with IRS. For this evaluation, the nasofibroscope is used.⁶ The priority in the treatment of IRS is the maintenance of the airway permeability as early as possible.^{7,20} However, it is observed that, although the improvement of the respiratory pattern is evident, infants develop hyper-secretiveness in the upper airways, requiring care for the maintenance of their permeability.¹³

The "impaired spontaneous ventilation" is defined as reduced energy reserves, resulting in the inability to maintain independent breathing that is adequate to sustain life.¹² In the present study, the prevalent defining characteristics were: dyspnea, increased use of accessory muscles and increased heart rate. Regarding the related factors, it prevailed to the respiratory muscle fatigue. All of these changes relate to the related symptomatology in the IRS triad.¹⁻²

The excessive use of the accessory muscles for a long time results in fatigue. In order to eliminate or reduce the respiratory distress, INF is used. The priority care relates to the hygiene of the cannula with saline, aspiration of the cannula and upper airways, as appropriate. Another intervention consists of monitoring through the pulse oximetry.¹⁰

Regarding feeding problems, the following nursing diagnoses were found: "Ineffective

breastfeeding", "Ineffective of the infant feeding pattern" and "Unbalanced nutrition: less than the bodily needs", according to the literature.¹³

"Ineffective breastfeeding" is defined as difficulty in offering milk to an infant or young child directly from the breasts, which may compromise the nutritional status of the infant/child.¹² This was observed in 75% of the participants of this study. The predominant defining characteristic was suction in the unsustained breast. Regarding the related factor, the oropharyngeal defect prevailed. These findings corroborate another study.¹³

The unsatisfactory sucking in children with IRS is caused by the respiratory obstruction that causes difficulty between the coordination of the suction, swallowing and breathing. In addition, the glossoptosis impairs tongue pre-emption required for the proper suctioning, and the cleft palate that causes less negative intraoral pressure required for and efficient suctioning.^{6,23-25}

A prospective study involving children with IRS showed that in the neonatal period, no children were breastfed, and only half of the children were fed a bottle, even partially in the first months, reflecting their feeding difficulty. However, at 8 months of age, most were weaned from the feeding tube and at 12 months of age, 85% were completely orally fed. This finding supports the inference that dietary problems cease or improve significantly and naturally at 12 months of age.²⁶

The diagnosis "Ineffective feeding standard of the baby" is defined by an infant's impaired ability to suck or coordinate the sucking/deglutition response, resulting in an inadequate oral nutrition for metabolic needs.¹² In the present study, the anatomical abnormality that incapacitates them from coordinating sucking, swallowing, and breathing was related. Similar results were evidenced in other studies.^{2,13,26}

The difficulty in breastfeeding by the oral route leads the child to be fed by the nasogastric tube. The exclusive breastfeeding is always encouraged. In case of oropharyngeal dysphagia, it is suggested to milk the milk that is offered through feeding tubes.²³

The nursing diagnosis of "Unbalanced Nutrition: less than the bodily needs" is defined as "insufficient intake of nutrients to meet the metabolic needs."¹² This was evidenced in 95% of the participants. The prevalent defining characteristics included: perceived inability of food

intake and insufficient muscle tone. Regarding the related factors, the inability to ingest food prevailed.

The decrease in the effectiveness of the genioglossus muscle in sustaining the tongue outside the pharynx causes its retro-positioning. The glossoptosis is accentuated by the micrognathia.¹⁻² Thus, for a proper suctioning, it is necessary to form negative intraoral pressure. In children with cleft palate this pressure is insufficient, interfering with the volume and flow of the liquid to be ingested, making the suction inefficient, and making the feeding process difficult or uncoordinated.²⁴⁻²⁵

In the treatment of eating difficulties in infants with IRS, feeding and hyper caloric diets are included.⁶ These consist of the stimulation of the non-nourishing suction through the use of pacifiers; massage to relax; manual support to support the jaw; long, soft bottle nozzle with increased hole to one millimeter; placement of the beak exactly on the tongue, symmetrical global posture and rhythmic movements of the beak in the oral cavity during nutritive sucking.²⁴ The feeding tube is withdrawn when the dietary intake corresponds to 70% of the volume recommended by age, with an average time of less than 30 minutes, without interurrences, such as gagging, cyanosis or coughing.^{6-7,24}

Research on dysphagia in children treated with nasopharyngeal intubation and feeding techniques found a reduction in the risk of aspiration after three weeks of treatment. Most children have developed the ability to be orally fed.²⁴

A prospective study pointed out that the tube feeding was used for a shorter time in children who received partial feeding by bottle, even if little, compared to those who did not. The conventional feeding benefits the child and gratifies the mother at a time when maternity is put to the test.²⁶

Another nursing diagnosis listed in this study was the "Risk of infection", defined as vulnerability to invasion and multiplication of pathogenic organisms that can compromise the health.¹² All the participants presented this diagnosis, whose prevalent risk factor was an invasive procedure.

Infants have an immature immune system, which makes them more susceptible to infectious agents. The fact that the infants are in a hospital setting and often are not adequately immunized, are the base for these factors.²⁷ The infection should be prevented with technical and

behavioral measures. It should be highlighted the hand-washing by health professionals, parents and caregivers between the interventions.²⁸

The diagnosis of "Risk of damage to skin integrity" is defined as vulnerability to changes in the epidermis and/or dermis, which may compromise health.¹² The prevalent risk factors were: extreme premature and mechanical factors. In contrast, the research pointed to the prevalence of only mechanical risk factors.²⁹

The fact that infants with IRS frequently require SNG and INF favors the risk of injury to the skin or mucosa due to the fixation and introduction of the device by professionals. A study indicated that infants are prone to this age-old diagnosis, fixation of catheters and probes, use of diapers and bony prominences.³⁰

Finally, we consider it important to emphasize the scarce literature on the accuracy of nursing diagnoses in infants with IRS, which limited the discussion of the results through comparisons, being a limitation of this research.

However, the benefits of this research are evident, since the identification of the nursing diagnoses and their respective defining characteristics and related risk factors is essential in the clinical practice to list the specific nursing interventions and direct them to the real needs of the clientele. It is emphasized that in accordance with the principle of decentralization of health services, these infants will be cared for in different locations. Thus, the present study may contribute to guiding the nursing care.

CONCLUSION

Infants with isolated Robin Sequence presented nursing diagnoses that related to respiratory, feeding, safety and comfort problems and favored the planning and implementation of nursing care while tracing a care profile.

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