ABSTRACT
Objective: to develop an expert system to support nurses for guiding children with Cystic Fibrosis. Method: methodological study that uses expert system based on classical logic for development of a model to support decision-making about the care of children with Cystic Fibrosis. Results: all newborns should be referred to perform the TNN; in case of negative result, the nurses follow with routine visits normally. Otherwise, they should investigate the family history of the child, neonatal intestinal obstruction, performing respiratory and nutritional assessment. Once detected any complications, they should send the child to the secondary or tertiary service; in all cases, guiding the search for social support. Conclusion: the model can contribute to a better direction to be followed by children with cystic fibrosis and their families. Descriptors: Technical Support For Decision; Cystic Fibrosis; Nursing; Primary Health Care.

RESUMO
Objetivo: elaborar um sistema especialista como suporte aos enfermeiros para direcionamento de crianças com Fibrose Cística. Método: estudo metodológico que utiliza o sistema especialista baseado em lógica clássica para elaboração de um modelo de apoio à tomada de decisão sobre a atenção à criança com Fibrose Cística. Resultados: todos os recém-nascidos devem ser encaminhados para realizar o TNN; em caso de resultado negativo, o enfermeiro segue com as consultas de puericultura normalmente; caso contrário, deverá investigar a história familiar da criança, obstrução intestinal neonatal, realizar avaliação respiratória e nutricional; uma vez detectada alguma complicação, deve-se encaminhar a criança ao serviço secundário ou terciário; para todos os casos, orientar a busca por apoio social. Conclusão: o modelo pode auxiliar no melhor direcionamento a ser seguido pela criança com fibrose cística e sua família. Descriptores: Técnicas de Apoio Para Decisão; Fibrose Cística; Enfermagem; Atenção Primária à Saúde.

ORIGINAL ARTICLE
SPECIALIST SYSTEM TO SUPPORT NURSES IN GUIDING CHILDREN WITH CYSTIC FIBROSIS
SISTEMA ESPECIALISTA PARA SUPORTE AO ENFERMEIRO NO DIRECIONAMENTO DE CRIANÇAS COM FIBROSE CÍSTICA

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INTRODUCTION

Cystic Fibrosis (CF) is a chronic genetic, autosomal, recessive disease. It results from mutations in a gene responsible for encoding the protein Cystic Fibrosis Transmembrane Conductance Regulator (CFTR) that acts regulating the balance between ions and water across the epithelium. It is an even incurable chronic disease affecting especially the white race individuals of both genders.1,3

CF clinical manifestations are linked to increases in viscosity and thickness of the mucous secretions, causing the obstruction of the exocrine glands ducts. This fact contributes to the emergence of three major complications: Chronic Obstructive Pulmonary Disease (COPD); and pancreatic insufficiency; secondary malnutrition.4,14

CF diagnosis in most cases is carried out when a child is between 4 to 7 years old and already presents a complication introduced above. Late diagnosis is a contributing factor to decreased survival of individuals affected by the disease.5

In this sense, the Neonatal Screening Test (TNN), known as “heel prick test”, enables to detect until the second month of life, to confirm the diagnosis of many diseases, including CF, increasing the possibility of survival of affected individuals and favoring subsidies for considerable improvements in the quality of life of individuals and their families.4,6 In this context, it is essential that care to this patient is offered in the three levels of care proposed by the Unified Health System (SUS) namely primary, secondary and tertiary levels.7,8

In primary care, the Family Health Team (ESF) is responsible by the children’s care, but it is the nursing professional that commonly performs child care consultation more closely. This is realized by periodical and systematic monitoring of children, growth and development surveillance, monitoring of immunization, food and hygiene guidelines for mothers, accident prevention and early detection of diseases, aimed at effective and appropriate intervention.9

However, in the context of primary care, the challenge for the professional nursing is a large and diverse demand for work, assigning to them the responsibility for carrying out activities involving situations whose solutions are complex, and needing following little routine protocols. Adding to this, many health professionals have difficulty working in what cannot be fully measured, that is, it depends on perception and use of soft and soft-hard technologies of care (approach, bond, expertise, well structured, normalized).10

Thus, the elaboration of a decision flowchart based on rules that support them in decision-making becomes very relevant. In this sense, the objective of this study is to develop an expert system as support to nurses to primary care for guiding and referral of children with Cystic Fibrosis.

METHOD

Methodological study that uses expert system based on classical logic for development of a model to support decision-making. It corresponds to a systematic method that sets aspects or stages to be considered by the decision maker (in this study, nurses), towards achieving a satisfactory outcome. It is a method of easy application and understanding.10

In this type of study, knowledge is represented through rules guiding to most appropriate decision-making every situation. Construction of the decision flowchart is done on the computer in an easily understandable format, using the logical operators “IF” and “THEN” that indicate the condition for rule activation and the decision to be made, respectively.11 In addition to these logical operators, connective “AND” and “OR” for chaining rules can be used. The connective “AND” says that all conditions must be met for that decision to be taken and the “OR” requires that at least one of the conditions is met.12

The results obtained in this study happened through two steps. The first one was the detection of the problem. While working in a Basic Health Unit (UBS), it was observed that many protocols are unfamiliar, complex and difficult to understand by the ESF professionals, such as the action on a case of cystic fibrosis. The second step was the formulation of rules that will help decision-making and had as manuals basis, protocols and recommendations of experts that support on the care of children with CF in the Basic Health Unit.1,7,13,14

In this sense, the decision flowchart was constituted as follows: “SE” (x) “THEN” (and), where (x) being the type of case brought by the child and (y) the action to be performed by nurses of primary care.

RESULTS AND DISCUSSION

As discussed above, CF is a genetic disorder that can be diagnosed early by the Newborn Screening Test. This test is established in Brazil since 2001 and should be performed, for free and compulsory, in all newborns, aiming
to research and early diagnosis of congenital diseases.\textsuperscript{4,15} Currently, NST is routinely performed in hospitals and referral hospitals at discharge from the newborn. However, in situations where the primary care nurse is faced with newborns who did not undergo the NST, his must direct the child to the referral service to conduct this test (Figure 1).

The Brazilian government established stages for the implementation of the National Neonatal Screening Program (PNTN) according to each state’s percentage of coverage, that is, states that have a 50% population coverage could triage hemoglobinopathies and states whose coverage reaches 70% the population is allowed to screening for Cystic Fibrosis.\textsuperscript{15}

Despite the TNN for CF not be a reality for all of the Brazilian states, the new demands that arise in health, notably by scientific and technological advances that at the time preventing infectious diseases through immunotherapy, also allow access the effective therapeutic methods for the treatment of chronic diseases, requiring that public policies are conceived and developed for this new population that is presented and that health professionals are able to act interdisciplinary, in situations that require intervention in both the individual and the family.\textsuperscript{14}

In situations where the child comes to UBS with the negative result of TNN for Cystic Fibrosis, the nurse should follow with routine visits, as recommended by the Ministry of Health (Figure 1).\textsuperscript{16} The childcare consultation is one of the activities ESF is fundamental to do preventing several diseases during the first years of life. Therefore, it is necessary that the first one is held, preferably, in the first month of life and the other at least eight times in a year.\textsuperscript{17} It is important that in the childcare consultation, the nurse allows that the meeting with the mother and the child happen in a confident way, to build a bond that lasts into adulthood.

The organs that depend on CFTR - lungs, pancreas, intestines, sweat glands and vas deferens - react differently to the deficit of this protein, causing major complications of CF.\textsuperscript{3} Among them, the most serious are: pancreatic insufficiency, causing bad absorption of nutrients and, consequently, weight loss and secondary malnutrition, requiring the nutritional assessment of children in care consultation (Figure 1); in the intestine due to a lack of water in the lumen, the fecal material becomes thick, contributing the subjective aspects that can affect the health-disease process of each individual and family.\textsuperscript{18}

Considering this discussion on the bond, is when the professional facing with a positive TNN for CF, and making appropriate referrals (Figure 1) should remain with the childcare consultations with the affected binomial, always paying attention to signs suggestive of complications from Cystic Fibrosis.

Figure 1. Expert System based on classical logic as support to nurses of primary care in the diagnosis and referral of children with cystic fibrosis.
to the occurrence of obstructive syndromes, hence the need for nurses to investigate intestinal obstruction in childcare consultation (Figure 1) and; respiratory evaluation (Figure 1) becomes important because lung disease is the leading cause of morbidity and mortality in CF, though not always be the most imminent sign of the disease. That is due to the impermeability of the apical membrane to chlorine, making it difficult to exit. Consequently, moisturization cell surface is compromised and there is a thickening of the pulmonary secretions, leading to obstruction of the airways and increased susceptibility to infections.

In health care, integrality becomes an essential principle for the conquest of “welfare” in this new concept of health. The reference and counter reference is one of the strategies to ensure universal, equitable and comprehensive health care since it provides dialogue between the various cores and professionals in the three performance levels of SUS. 19

Therefore, the network should be structured so that each service complete the action of the other through organized and agreed mechanisms. 19 However, this tool still is far from the reality of SUS, requiring structural changes and in the traditional way of professional practice in health.

Given this reality, to identify complications resulting from CF childcare consultation the nursing professional shall forward in a responsible manner and through dialogue, the child and his family to the secondary or tertiary service according to regional pacts (Figure 1).

In addition to coordination between health care levels, the nursing professional of primary care must know the network of support to the binomial at the local level (neighbors, friends, family, church), encouraging the search for social support. 17,20

In Brazil, in most states, there are associations that help children with CF and their families. In the state of Paraíba is the Paraíba Association of Parents and Patients with Cystic Fibrosis (ASPAFIC) that acts favoring the sharing of experiences among members and allowing greater confidence of juridical power and social legitimacy in the development of actions aimed the right to children’s lives. 21 They also assume the educational role of collaborating with studies, research and assistance to patients with this pathology. 1, 19

In this sense, the nursing professional must guide the family to seek support from the Association of their state (Figure 1), in view of its importance to face CF treatment and the difficulties inherent to illness in childhood.

**CONCLUSION**

Chronic disease in childhood and all its psychobiological implications are able to interfere in the daily lives of children and their family microsystem. This is the center of health actions undertaken by the Primary care, more precisely, by the Family Health Strategy. The performance of primary care nursing professional with the child with Cystic Fibrosis and his family needs to happen effectively, through childcare, cozy, human, confident and qualified way.

The child with CF live with constant hospitalizations, which can be understood by the family, as a substitute service monitoring done by the nurse on a routine visit. In this sense, the nursing professional must know the reality of their territory and act with ethics, responsibility and interdisciplinary, demystifying inadequate concepts and valuing approach these clients with the service offered at primary level.

Knowing the need for such monitoring and the great work demands of nurses who work in primary care, the expert system based on classical logic, developed in this study can provide this professional core decision support and hence solving in the best direction to be followed by children with CF and their families. Thus, there are expectations of diagnosis and early treatment, favoring the quality of life of this population. In addition, the arising result of this research is also an educational and innovative product in the health field, as it allows the dissemination of synthesized way protocols, easy to understand and apply.

It is important to highlight that the logical model facilitates the work of professionals, but does not replace care practices that involve the subjectivity of the subject and that are still needed for the expansion of care to the binomial studied here.

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