INTEGRATIVE LITERATURE REVIEW ARTICLE

OUTPATIENT NURSING CARE FOR CYSTIC FIBROSIS PATIENTS ASSISTÊNCIA DE ENFERMAGEM NO AMBULATÓRIO AO PACIENTE COM FIBROSE CÍSTICA ASISTENCIA DE ENFERMERÍA EN EL AMBULATORIO AL PACIENTE CON FIBROSIS CÍSTICA

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ABSTRACT

Objective: to analyze the scientific evidence of outpatient nursing care for patients with cystic fibrosis. **Method:** this is a descriptive, bibliographic study, of the type integrative literature review, carried out with 14 articles available online, published from 2013 to 2017 in MEDLINE database, CINAHL, LILACS, BDENF, IBECs, and WEB OF SCIENCE. Data were analyzed according to the Content Analysis technique and were presented in figures. **Results:** the findings pointed out that the outpatient nursing care for cystic fibrosis patients is still incipient. There are difficulties in drug adherence, pain control, lack of involvement of families in the treatment, and decreased quality of life and comfort of patients. **Conclusion:** international articles mainly published in English and with level of evidence VI were observed, and there were more productions about cystic fibrosis in the pediatric area. It is understood that the themes that will contribute to the advancement of nursing in future studies are: medication adherence for treatment of cystic fibrosis; family-centered care; psychosocial support to clients and families; quality of life; and promotion of comfort; and pain relief. **Descriptors:** Cystic Fibrosis; Nursing care; Chronic Disease; Pneumology; Genetics; Nursing.

RESUMO

Objetivo: analisar as evidências científicas acerca dos cuidados de Enfermagem no âmbito ambulatorial voltados aos pacientes com fibrose cística. **Método:** trata-se de estudo descritivo, bibliográfico, tipo revisão integrativa, com 14 artigos disponíveis *on-line*, no período de 2013 a 2017, nas bases de dados MEDLINE, CINAHL, LILACS, BDENF, IBECs, *WEB OF SCIENCE*, analisados pela Técnica de Análise de Conteúdo, e apresentados em forma de figuras. **Resultados:** avalia-se que a assistência de Enfermagem no ambulatório ao paciente com fibrose cística ainda é incipiente; com isto, há dificuldades na adesão medicamentosa, controle da dor, falta de envolvimento da família no tratamento, diminuição da qualidade de vida e conforto do paciente. **Conclusão:** observou-se uma literatura internacional constituída, majoritariamente, por produções em inglês, com nível de evidência VI, e percebeu-se que há mais produções sobre a fibrose cística na área pediátrica. Entende-se que os temas que contribuirão para o avanço da Enfermagem em estudos futuros serão: a adesão medicamentosa ao tratamento para a fibrose cística; os cuidados centrados na família; o apoio psicossocial ao cliente e família; a qualidade de vida; a promoção do conforto e o alívio da dor. **Descritores:** Fibrose Cística; Cuidados de Enfermagem; Doença Crônica; Pneumologia; Genética; Enfermagem.

RESUMEN

Objetivo: analizar las evidencias científicas acerca de los cuidados de Enfermería en el ámbito ambulatorial dirigidos a los pacientes con fibrosis cística. **Método:** se trata de un estudio, descriptivo, bibliográfico, tipo revisión integradora de la literatura, con 14 artículos disponibles *on-line*, en el período de 2013 a 2017, en las bases de datos MEDLINE, CINAHL, LILACS, BDENF, IBECs, *WEB OF SCIENCE*, estudiados según el Análisis de Contenido y presentados en forma de figuras. **Resultados:** se evalúa que la asistencia de Enfermería en el ambulatorio al paciente con fibrosis cística aún es incipiente, con eso hay dificultades en la adhesión medicamentosa, control del dolor, falta de involucramiento de la familia en el tratamiento, disminución de la calidad de vida y confort del paciente. **Conclusión:** se observó una literatura internacional constituída mayoritariamente por producciones en inglés, con nivel de evidencia VI, y se percibe que hay más producciones sobre la fibrosis cística en el área pediátrica. Se entiende que los temas que contribuirán para el avance de la Enfermería en estudios futuros serán la adhesión medicamentosa al tratamiento para la fibrosis cística; los cuidados centrados en la familia; el apoyo psicossocial al cliente y familia; la calidad de vida; la promoción del confort; y el alivio del dolor. **Descriptores:** Fibrosis Quística; Atención de Enfermería; Enfermedad Crónica; Neumología; Genética; Enfermería.

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INTRODUCTION

It is recognized that cystic fibrosis (CF) or mucoviscidosis, as it is popularly known, is a serious hereditary disease determined by an autosomal recessive inheritance pattern that especially affects the lungs and pancreas in an obstructive process caused by increasing mucus viscosity. It is noteworthy that, in the lungs, the increased viscosity blocks the airways, favoring bacterial proliferation (especially *pseudomonas* and *staphylococci*), which leads to chronic infection and lung injury, with loss of capacity and elasticity, ultimately resulting in damage and death due to respiratory dysfunction. In turn, the obstruction of ducts in the pancreas by thick secretion causes loss of digestive enzymes, leading to malnutrition.¹

In Brazil, it is estimated that the incidence of cystic fibrosis is 1: 7576 live births. However, there are variations between regions, with higher values in the states of the South.² It is known that CF is detectable as early as in neonatal screening, in the pre-symptomatic phase, in all live births, and is one of the main pulmonary diseases of childhood. In Brazil, it has an approximate incidence of 1:10 thousand live births.³

It is known that nurses in the outpatient care have to be especially attentive to pulmonary symptoms and they must guide patients or family members about the risk factors associated with the respiratory infections in order to avoid or reduce complications. Signs and symptoms of the disease in the airways are explained to patients and their families, and the need for adequate water and nutritional intake is stimulated. It is essential to explain any and all procedure in order to reduce the patients' anxiety and surprise to the new, reducing the fear of the unknown.⁴

It is pointed out that nurses must pay attention to the biopsychosocial condition of patients so as to provide care in an individualized manner. It is also important that the professionals develop new skills to care for and guide about the needs and expectations of cystic fibrosis patients and their families. It is argued, in this sense, that nurses develop an attitude of commitment to provide care in a humanized way, based on a relationship of help and compassion.⁴

OBJECTIVE

- To analyze the scientific evidence about nursing care in the outpatient context for cystic fibrosis patients.

METHOD

This is a quantitative, descriptive, bibliographical study, an integrative literature review that allows the identification, analysis and synthesis of knowledge addressed in independent studies about a given subject in order to point the existing gaps. The following steps were taken to prepare this integrative review: definition of the hypothesis and objectives of the integrative review; establishment of criteria for inclusion and exclusion of articles (sample selection); definition of information to be extracted from the selected articles; analysis of results; discussion and presentation of results and synthesis of knowledge.⁵

The articles were surveyed by two independent reviewers in April and May 2018. A protocol of search and revision of articles was used in the study, covering the identification of the theme and selection of the research question: "What are the outpatient nursing care measures used for patients with cystic fibrosis?".

The content was searched through online access to important database systems in the context of health: Medical Literature Analysis and Online Retrieval System (MEDLINE, via EBSCO), Cumulative Index to Nursing and Allied Health Literature (CINAHL), Latin American and Caribbean Literature in Health Sciences (LILACS), IBECs, WEB OF SCIENCE and PubMed. The inclusion criteria were: articles published in English, Spanish or Portuguese, between the period 2013 and 2017, and available in full length. We excluded research without subsidies for nursing and which did not explicitly address nursing care to cystic fibrosis.

Health Sciences Descriptors (DeCS) and combinations were associated in a strategy ("cloud of DeCS combinations") were associated: "cystic fibrosis", "nursing care" and "nursing", linked by the Boolean operator *and*, used with appropriate selection of "TITLE, ABSTRACT AND SUBJECT". After final composition of the database, the study was guided by the Content Analysis technique to discuss the content of the articles.

Seven levels of evidence for clinical practice were explored and described in this study: level I for systematic reviews or meta-analyses; level II for randomized controlled trials; level III for non-randomized controlled trials; level IV for case control studies or cohort studies; level V for qualitative meta-syntheses; level VI for qualitative studies; and level VII for expert

opinions.⁶ The PRISMA recommendations shown in Figure 1 were used to select the publications to be included in the study.

The ethical precepts involved in the analysis and dissemination of research data were respected because this was a sort of documentary study whose data come from public character.

Figure 1. Flowchart Adapted from the PRISMA 2009 model used in the selection of studies. Belém, PA, Brazil, 2018.

RESULTS

A total of 219 articles were found, of which 18 referred to nursing care for patients with cystic fibrosis. Four were excluded because they were repeated in different databases. Fourteen articles pertinent to the objectives were selected for analysis. The distribution of

the articles by database was: MEDLINE (n = 9), PubMed (n = 4), and WEB OF SCIENCE (n = 1). It should be noted that no articles meeting the inclusion criteria were found in the databases BDNF, CINAHL, LILACS and IBECs. Most articles had level of evidence VI, that is, they were qualitative and/or descriptive articles.

Title and Identification Code	Authorship/Source/Year/ Level of evidence	Objective	Recommendations/ Conclusions
Evaluation of pain, dyspnea, and goals of care among adults with cystic fibrosis: a comprehensive palliative care survey/L1	Chen E, Killen KM, Peterson SJ, Saulitis AK, Balk RA/MEDLINE/2017/VI	To describe the perceptions of patients of pain, dyspnea, and planning of advanced care.	Pain and dyspnea are common among adults with cystic fibrosis. Few patients had an updated previous directive, but most were open to discuss end-of-life care issues. The results of this unicentric study may not represent the entire population; therefore, a multicentric investigation is yet needed.
Protocol for a study of the psychosocial determinants of health in early childhood among children with cystic fibrosis/L2	Douglas T, Jordan B, Priddis L, Anderson V, Sheeran J, Kane RT et al/MEDLINE/2014/IV	To investigate the causal associations between family relationships, family functioning, social circumstances, and health outcomes in children with cystic fibrosis.	This was the first study to investigate the causal effect of psychosocial functioning, kinship, and attachment on measures of physical health outcomes in children with cystic fibrosis.
Parental experience of information and education processes following diagnosis of their infant with cystic fibrosis via newborn screening/L3	Jessup M, Douglas T, Priddis L, Branch-Smith C, Shields L/ MEDLINE/2015/VI	To understand the initial parental experiences of education after the diagnosis of cystic fibrosis in their babies during neonatal screening.	Effective education informs the individuals, initiates them into the world of cystic fibrosis, helps them establish a care regimen, integrates them with the care team, and empowers them to achieve this extraordinary and unexpected level of parental care.
Chasing Zero: Increasing infection control compliance on an inpatient cystic fibrosis unit/L4	Johnson S, McNeal M, Polineni D, Burger S/MEDLINE/2017/IV	To assess the adherence of the staff of the inpatient unit to current guidelines of the Cystic Fibrosis Foundation for appropriate use of PPE, adherence of staff to organizational standards regarding the use of Safe Zone, and whether educational interventions improved with the effective infection prevention.	To improve infection control and compliance with PPE, which are directly linked to the prevention of transmission and acquisition of respiratory pathogens. The interventions in this project promote and facilitate the best infection prevention practices in the population with cystic fibrosis.
Advance care planning in adolescents with cystic fibrosis: a quality improvement project/L5	Kazmerski TM, Weiner DJ, Matisko J, Schachner D, Lerch W, May C et al/MEDLINE/2016/IV	To present advanced care planning to patients and families, improve caregiver understanding, and assess patient attitudes and preferences regarding advanced care.	Adolescents with advanced cystic fibrosis disease felt that advanced care planning was a positive, non-detrimental experience. Cystic fibrosis caregivers valued advanced care planning, but they wanted more training.
They know it's safe - they know what to expect from that face: perceptions towards a cognitive-behavioural counselling programme among caregivers of children with cystic fibrosis/L6	Moola FJ, Henry LAV, Huynh E, Stacey JA, Faulkner GEJ/ MEDLINE/2016/VI	To explore the experiences of eight caregivers who cared for children with cystic fibrosis in an eight-week cognitive-behavioral counseling program at a children's hospital in Winnipeg, Canada.	From this evidence-based cognitive-behavioral counseling program, views on the complex psychosocial lives of the cystic fibrosis community are discussed within the context of the literature. Integrating cognitive behavioral counseling into routine clinical care for CF should be considered as a method to improve the capacity to care for the CF community and should be advocated by nurses.
Caregiver coping, mental health and child problem behaviours in cystic fibrosis: a cross-sectional study/L7	Sheehan J, Hiscock H, Massie J, Jaffe A, Hay M/MEDLINE/2013/IV	To identify, through a principal component analysis, the coping strategies used by Australian caregivers of children with CF and assess the relationship between derived coping components, mental	Defensive coping strategies are correlated with the caregiver's mental health and the child's problematic behaviors. Intervening with caregiver coping can be a way to improve both the caregiver's mental health and the problematic behaviors of children with cystic fibrosis.

		health symptoms of the caregiver, and problematic behaviors related and non-related to the treatment of the child.	
Family-centred care in cystic fibrosis: a pilot study in North Queensland, Australia/L8	Smyte W, Abernethy G, Jessup M, Douglas T, Shields L/MEDLINE/2017/IV	To examine the perceptions of family-centered care of parents of children with cystic fibrosis and their caregivers; to test tools in a regional population.	Evidence can be translated into practice, and consequent improvements in care provision in the application of results will mean better clinical outcomes for children, along with psychosocial outcomes for families and the community.
How holistic nursing can enhance the quality of life of children with cystic fibrosis/L9	Toiton K, Hunt J/MEDLINE/2016/VI	To discuss, based on a case study, the effects of cystic fibrosis on a patient and his family and how this affects his quality of life and well-being.	Positive and negative coping strategies may exist within a family unit; it is, thus, essential to be attentive to all family members whenever possible and ensure that each of them receives appropriate holistic nursing care to improve his well-being.
Descriptions of the pain experience in adults and adolescents with cystic fibrosis/L10	Allgood SJ, Kozachik S, Alexander KA, Thaxton A, Vera M/PUBMED/2017/VI	To explore descriptions reported by the patient about the pain experience among adolescents and adults living with cystic fibrosis.	Adolescents experience episodic pain, feel supported in their pain management, and run the risk of loss in social functioning as a result of the emotional impact of social events they miss. Adults experience persistent pain, recognize a distinct emotional pain associated with cystic fibrosis, and feel stigmatized and unsupported in their attempts to seek treatment for pain.
Guidelines for the transition from child to adult cystic fibrosis care/L11	Al-Yateem/PUBMED/2013/VI	To develop relevant and feasible guidelines for transition care, based on the interested parties perspectives.	The proposed guidelines reflect essential care elements for a safe transition of the child to adult health services. They will, in some way, meet the diverse needs of young people living with a chronic disease that limits life.
An exploration of partnership through interactions between young 'expert' patients with cystic fibrosis and healthcare professionals/L12	MacDonald K, Irvine L, Smith MC/PUBMED/2015/VI	To explore how the young "specialist patients" who deal with cystic fibrosis and the health professionals with whom they interact perceive partnerships and negotiate care.	The consultation model can be a barrier to person-centered care. Health professionals show leniency in the negotiations, but they do not always trust patients' accounts. The term "specialist patient" is unpopular and remains contested.
Transferring young people with cystic fibrosis to adult care/L13	Tierney et al/PUBMED/VI	To explore activities, interactions and behaviors during the first consultation in adults for young people with cystic fibrosis.	A modest set of goals for the first consultation should be set to allow young people to retain information when introduced to a new service.
Communication, comfort, and closure for the patient with cystic fibrosis at the end of life/L14	Price DM, Knotts SE/Web of Science/2017/VI	To describe the critical role of the nurse at the bedside in the case of a young patient with cystic fibrosis and promote a dignified and peaceful death.	In the terminal care of patients with cystic fibrosis, the role of the nurse at the bedside is critical to providing continuous and interdisciplinary care in order to promote a good death and ensure that the wishes of the patient and the family are respected at the end of life.

Figure 2. Distribution of the articles found related to Nursing care and cystic fibrosis: titles and identification code; authorship/source/year/level of evidence; goal; recommendations/conclusions. Belém (PA), Brazil, 2018.

DISCUSSION

Patients with cystic fibrosis have shown to develop recurrent respiratory tract infections. Intravenous antibiotic treatment is indicated

for both moderate and severe respiratory exacerbations, and cyclic treatment is indicated for patients chronically colonized by *Pseudomonas aeruginosa* and, in the latter case, adverse effects and increased bacterial

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resistance are more frequent. The patient is guided both in medication adherence in cases of infection and prevention of cystic fibrosis, such as malnutrition and decrease of fat-soluble vitamins (K, E, D and A). Hospital infection levels are reduced through measures such as hand washing, use of personal protective equipment, and hygiene of materials, and asepsis in procedures; nurses are responsible for supervising these measures.⁷⁻⁸

Investments are currently placed on family-centered nursing care, especially in Pediatric Nursing, and this is reflected in the care of patients with cystic fibrosis, since most patients discover the disease as early as in childhood. It is noted that family-centered care is based on a set of fundamental principles: dignity and respect; information sharing; participation and collaboration; context-based negotiation and care. In this proposal, the professional-patient interaction is increased, favoring the quality of care and, consequently, more effective results and a shorter recovery time.⁹⁻¹¹

It is important to emphasize, in relation to family-centered care, that it is important for nurses to promote psychosocial support to patients and their families, since caregivers and people with cystic fibrosis have high rates of psychosocial morbidities such as depression, anxiety, difficulties of adherence and adaptation to the new condition and uncertainty of prognosis.¹¹⁻⁶

It is pointed out that patients with cystic fibrosis face a high burden of symptoms, tending to a low quality of life, mainly due to the excess of medications, restrictive diet, and recurrent respiratory infections that are part of the routine of these patients, reflected in the education, which is interrupted by recurrent hospital admissions. The nurses become transforming agents of this situation, giving the correct guidance in outpatient nursing consultations regarding the treatment of the patients.¹⁷⁻⁹

The main symptoms of discomfort in patients with cystic fibrosis include shortness of breath, difficulty in expectoration, and chest pain. Chest discomfort in cystic fibrosis patients is related to frequent episodes of cough, resulting in stress on the chest wall. Pain relief, relaxation and comfort are promoted through alternative therapies such as massage, as well as through the administration of analgesics according to the frequency and intensity of

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pain, tasks performed by nurses.²⁰⁻² This study was limited by the non-use of all databases of the health area.

CONCLUSION

International literature composed mainly of English productions with level of evidence VI was observed and it was noticed that there are more productions on cystic fibrosis in the pediatric area. It is understood that the themes that will contribute to the advancement of nursing in future studies are: medication adherence for treatment of cystic fibrosis; family-centered care; psychosocial support to clients and families; quality of life; promotion of comfort; and pain relief.

REFERENCES

1. Ministério da Saúde (BR), Secretaria de Atenção à Saúde, Departamento de Atenção Especializada e Temática. Triagem neonatal biológica: manual técnico [Internet]. Brasília: Ministério da Saúde;2016. [cited 2018 June 21]. Available from: http://bvsms.saude.gov.br/bvs/publicacoes/triagem_neonatal_biologica_manual_tecnico.pdf
2. Athanazio RA, Silva Filho LVRF, Vergara AA, Ribeiro AF, Riedi CA, Procionoy EDFA, et al. Brazilian guidelines for the diagnosis and treatment of cystic fibrosis. J Bras Pneumol. 2017 May/June;43(3):219-45. DOI: <https://doi.org/10.1590/S1806-37562017000000065>
3. Monte LFV, Goya A. Diretriz Interprofissional de Atenção à Criança e ao Adolescente com Fibrose Cística [Internet]. Brasília: HCB;2016. [cited 2018 June 21]. Available from: http://www.hcb.org.br/arquivos/downloads/hcb2016_manual_10_cadernopaciente_vdigital.pdf
4. Mariano T, Conde CR. Child to the nurse's assistance with cystic fibrosis. Revista Uningá [Internet]. 2017 Jan/Mar [cited 2018 Aug 15];52(1):144-50. Available from: https://www.mastereditora.com.br/periodico/20170504_222156.pdf
5. Soares CB, Hoga LAK, Peduzzi M, Sangaleti C, Yonekura T, Silva DRAD. Integrative review: concepts and methods used in nursing. Rev esc enferm USP. 2014 Apr;48(2):335-45. DOI: <http://dx.doi.org/10.1590/S0080-6234201400002000020>
6. Melnyk BM, Fineout-Overholt E, Stillwell SB, Williamson KM. Evidence-based in practice: step by step. Am J Nurs. 2010 Jan;110(5): 41-7. DOI: <https://doi.org/10.1097/01.NAJ.0000366056.06605.d2>
7. Girón RM, Martínez A, Máiz L, Salcedo A, Beltrán B, Martínez MT, et al. Tratamiento antibiótico intravenoso domiciliario en las unidades de fibrosis quística de la Comunidad de Madrid. Med

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clin (Barc). 2004;122(17):648-52. DOI: [https://doi.org/10.1016/S0025-7753\(04\)74341-9](https://doi.org/10.1016/S0025-7753(04)74341-9)

8. Johnson S, McNeal M, Mermis J, Polineni D, Burger S. Chasing Zero: Increasing infection control compliance on an inpatient cystic fibrosis unit. *J Nurs Care Qual.* 2018 Jan;33(1): 67-71. DOI: <https://doi.org/10.1097/NCQ.0000000000000269>

9. MacDonald K, Irvine L, Smith MC. An exploration of partnership through interactions between young 'expert' patients with cystic fibrosis and healthcare professionals. *J Clin Nurs.* 2015 Dec;24(23-24):3528-37. DOI: <https://doi.org/10.1111/jocn.13021>

10. Smyth W, Abernethy G, Jessup M, Douglas T, Shields L. Family-centred care in cystic fibrosis: a pilot study in North Queensland, Australia. *Nursing Open.* 2017;4(3):168-73. DOI: <https://doi.org/10.1002/nop2.84>

11. Douglas T, Jordan B, Priddis L, Anderson V, Sheehan J, Kane RT, Massie J, et al. Protocol for a study of the psychosocial determinants of health in early childhood among children with cystic fibrosis. *J Adv Nurs.* 2015 July;71(7):1704-16. DOI: <https://doi.org/10.1111/jan.12621>

12. Moola FJ, Henry LA, Huynh E, Stacey JA, Faulkner GE. They know it's safe - they know what to expect from that face: perceptions towards a cognitive-behavioural counselling programme among caregivers of children with cystic fibrosis. *J Clin Nurs* 2016 Oct;26(19-20):2932-43. DOI: <https://doi.org/10.1111/jocn.13622>

13. Sheehan J, Hiscock H, Massie J, Jaffe A, Hay M. Caregiver coping, mental health and child problem behaviours in cystic fibrosis: a cross-sectional study. *Int J Behav Med.* 2014 Apr;21(2):211-20. DOI: <https://doi.org/10.1007/s12529-013-9289-y>

14. Tierney S, Kirk S, Deaton C. Transferring young people with cystic fibrosis to adult care. *Nurs stand.* 2015;30 (16-18):41-7. DOI: <https://doi.org/10.7748/ns.30.16.41.s45>

15. Al-Yateem N. Guidelines for the transition from child to adult cystic fibrosis care. *Nursing children and young people.* Stand Rio 2013 Dec.34. DOI: [10.7748/ns.30.16.41.s45](https://doi.org/10.7748/ns.30.16.41.s45)

16. Quittner AL, Abbott J, Georgiopoulos AM, Goldbeck L, Smith B, Hempstead SE et al. International Committee on Mental Health in Cystic Fibrosis: Cystic Fibrosis Foundation and European Cystic Fibrosis Society consensus statements for screening and treating depression and anxiety. *Thorax.* 2016 Jan;71(1):26-34. DOI: <https://doi.org/10.1136/thoraxjnl-2015-207488>

17. Kazmerski TM, Weiner DJ, Matisko J, Schachner D, Lerch W, May C, et al. Advance care planning in adolescents with cystic fibrosis: a quality improvement project. *Pediatr Pulmonol.* 2016 Dec;51(12):1304-10. DOI: <https://doi.org/10.1002/ppul.23559>

18. Jessup M, Douglas T, Priddis L, Branch-Smith C, Shields L, AREST-CF. Parental experience of information and education processes following diagnosis of their infant with cystic fibrosis via

Outpatient nursing care for cystic fibrosis...

newborn screening. *J Pediatr Nurs.* 2016 May/June;31(3) e233-41. DOI:

<https://doi.org/10.1016/j.pedn.2015.11.010>

19. Tointon K, Hunt JA. How holistic nursing can enhance the quality of life of children with cystic fibrosis. *Nurs Child Young People.* 2016 Oct;28(8):22-5. DOI:

<https://doi.org/10.7748/ncyp.2016.e753>

20. Chen E, Killeen KM, Peterson SJ, Saulitis AK, Balk RA. Evaluation of pain, dyspnea, and goals of care among adults with cystic fibrosis: a comprehensive palliative care survey. *Am J Hosp Palliat Care.* 2017 May;34(4):347-52. DOI: <https://doi.org/10.1177/1049909116629135>

21. Allgood SJ, Kozachik S, Alexander KA, Thaxton A, Vera M, Lechtzin N. Descriptions of the pain experience in adults and adolescents with cystic fibrosis. *Pain Manag Nurs.* 2018 Aug;19(4):340-7. DOI: <https://doi.org/10.1016/j.pmn.2017.11.011>

22. Price DM, Knotts SE. Communication, comfort, and closure for the patient with cystic fibrosis at the end of life. *J Hosp Palliat Nurs.* 2017 Aug;19(4):298-302. DOI:

<https://doi.org/10.1097/NJH.0000000000000362>

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