FT12

Kistik Fibrozisli Çocuğun Bakımda Hemşiresinin Rolü

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Kistik fibrozis (KF), otozomal resesif geçişli, ekzokrin salgı bezlerinde fonksiyon bozukluğu ile karakterize, birçok sistemi tutan kronik bir hastalıktır. KF'de beklenen yaşam süresinin uzatılması ve hastalığın iyi prognoz göstermesinde multidisipliner bakım yaklaşımlarının tercih edilmesi önemli bir faktördür. KF'te hemşirelik bakımının amacı; çocuğun solunum fonksiyonlarının artırılması ve korunması, optimal düzeyde beslenmenin sağlanması, çocuğun yaşına uygun büyüme gelişmesinin sağlanması ve ebeveynlerin psikososyal açıdan desteklenmesidir. KF'nin yaşamı tehdit eden bir hastalık olması, sık hastaneye yatışlar, morbite riskinin yüksekliği, yaşanılan ekonomik ve sosyal sorunlar açısından çocuk hemşireleri ailenin yaşadığı sorunların farkında olmalı ve danışmanlık hizmeti vererek aileleri desteklemelidirler. Hemşirelik girişimleri ile desteklenen KFli çocukların mortalite ve morbidite oranları üzerine olumlu etkilerinin olduğunu göstermektedir. KF'li çocuk ve ebeveynlerine yönelik uygulanan planlı hastalık yönetimi eğitim girişimi ve aile güçlendirme programları, ebeveynlerin hastalık yönetimi becerisini artırmaktadır. Ebeveynlerin hastalık yönetimine ilişkin bilgi düzeylerinin artırılması, sorularının yanıtlanması ve ebeveynlerin kararlara katılımının sağlanması KF'li çocukların yaşam kalitelerini ve sürelerini artırıcı etkisi vardır. Hasta ve ebeveynin var olan potansiyellerinin geliştirilmesi ve yasal haklarının korunması çocuk hemşirelerinin savunucu rolllerinden bir tanesidir. Hemşirelerin KF'de bakıma ilişkin deneyimlerini ebeveynler ile paylaşmaları bakım kalitesinin geliştirilmesinde ve ebeveynin yaşadığı psikososyal sorunların azaltılmasında etkili bir girişimdir. KF'li adölesanlar ve ebeveynlerin bakım ihtiyaçlarının belirlenmesi ve hastalığın günlük yaşama adaptasyonunun sağlanmasında hemşirelik eğitimi önemli bir role sahiptir.

Anahtar Kelimeler: *Hemşire, Kistik Fibrozis, Çocuk, Bakım*

The Role of Nurses in the Care of a Child with Cystic Fibrosis

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Cystic fibrosis (CF) is an autosomal recessive chronic disease characterized by exocrine gland dysfunction. It affects many systems in the body. Multidisciplinary care approaches are an important factor in prolonging life expectancy and correct prognosis in CF. The aim of nursing care in CF is to increase and maintain the child's respiratory functions, to ensure optimal nutrition, to promote the growth of the child appropriate for his age, and to psychologically support the parents. Since CF is a life-threatening disease and it entails frequent hospitalization, high morbidity risk, and economic and social problems, pediatric nurses should be aware of the problems experienced by families and support them by providing counseling. Studies have shown that the mortality and morbidity rates of children with CF are positively affected by nursing interventions. Planned disease management training initiatives and family empowerment programs for children with CF and their parents increase the disease management skills of parents. Increasing the knowledge level of parents about disease management, answering their questions and ensuring the participation of parents in the decision making process increase the life quality and life expectancy of children with CF. Developing the existing potential of the patient and parents and protecting their legal rights are advocacy

roles of pediatric nurses. As nurses share their care experiences with parents, the quality of care improves and the psychosocial problems experienced by parents decrease. Nursing education plays an important role in determining the care needs of adolescents with CF and their parents and ensuring the adaptation of the patients to daily life.

***Keywords**: Nurse, Cystic fibrosis, Child, Care*

The Role of Nurses in the Care of a Child with Cystic Fibrosis

Cystic fibrosis (CF) is a complex, progressive, systemic and autosomal recessive disease characterized by exocrine secretory gland dysfunction. It involves many systems and is life-threatening (Hay et al., 2013, Yara et al., 2013). The frequency of the disease is 1/2000-3500 and it varies from country to country. Although the incidence of the disease was determined to be 1/3000 in limited number of studies conducted in Turkey, it is thought to be higher given that kin marriage is frequent in Turkey (SB, 2017).

Morbidity and mortality in CF are caused by bronchial obstruction and stasis in the lungs, chronic infection, inflammation, fibrosis, bronchiectasis and cystic dilatation. With a multidisciplinary approach to neonatal screening, care, and intensive symptomatic treatment, prognosis has improved dramatically over the past decade and thus, life expectancy has increased (Fajac & Wainwright, 2017).

Pediatric nurses who are the members of multidisciplinary care approach in CF actively advocate for improving the potential of the patient and his/her family and protecting their legal rights. Improving the life quality of the child with CF is the basic building block of nursing care at every stage from daily life to school experience and to the death of the patient. Nursing education topics in CF are nebulization practices and clearing the airways, hygiene, antibiotics treatment, long-term oxygen treatment, noninvasive ventilation, nutrition, psychosocial support for parents and adolescents, genetic counseling, and end-of-life care (Reisinho & Gomes, 2016,; Koeller & Meyer, 2016).

Antibiotics, mucolytics and bronchodilator drugs administered to the child with CF constitute the medical part of the treatment given for the preservation of pulmonary functions. Lifelong medical treatment is a source of anxiety for children and parents. In order to prevent the development of complications, pediatric nurses are responsible for improving the quality of life of the child during home care and educating parents about disease management (Çavuoğlu, 2013; Yara et al., 2013).

As far as hygiene is concerned, nebuliser applications, cleaning and disinfection, CF pathogens and the colonization of other respiratory equipment used at home, which all constitute a large part of the treatment, are the recommended training topics for families. Decreased frequency of disinfection is associated with the recovery of microorganisms on nebulizers. To encourage the cleaning and disinfection of nebulizers used at home after each use as recommended by CF care guidelines, trainings should be repeated at specific time periods (Castallani et al., 2018; Murray et al., 2019).

Pediatric nurses should be aware of the problems experienced by families as CF is a life-threatening disease, entails frequent hospitalizations, carries high morbidity risk, and leads to economic and social problems. Pediatric nurses should give counseling to the parents and support them in order to help them adapt to the disease (Torüner & Büyükgönenç, 2013; Nierengarten, 2017). It is reported that the counseling given to parents by nurses about the problems they experience during the care process is an effective method in improving the quality of care (Moola et al., 2016).

In a systematic review of eight articles examining nursing interventions in the care of children with CF, Reisinho and Gomes (2016) revealed that nursing interventions play an important role in identifying the care needs of the children with CF and their parents and in developing a strong adaptation to the disease. Reisinho and Gomes define the nursing interventions that are effective in normalizing daily life in CF as identifying educational needs for different age periods and educating parents, ensuring adaptation during the pre and post-hospitalization process, determining the educational needs of children and parents before and after lung transplantation, and providing emotional and psychological support. Evidence-based care guidelines, developed by pediatric nurses, have been reported to be effective in improving care at home, nutrition, nebulization therapy, oxygen therapy, daily life, school process and quality of life (Reisinho & Gomes, 2016). A qualitative study conducted with children with CF and their parents using a family-centered care model revealed that the communication developed with the support, respect and cooperation of the parents with the health personnel and especially the nurse group strengthens the parents (Smyth et al., 2017).

Hypertonic solutions, bronchodilators and antibiotics that need to be applied with nebulizers in CF include a treatment protocol that continues in daily life. This situation restricts participation in school activities and reduces compliance to treatment especially in school-age children and adolescents. The information and guidance provided by nurses as a member of the CF team play a significant role in improving the adaptation to the disease and reducing the treatment-related difficulties experienced by the children with CF and their parents (Tointon, K & Hunt, J., 2016; Gathercole, K., 2019).

In chronic illnesses or disabilities, the young or young adults experience numerous difficulties in the transition process from pediatric health care to adult health care. Pediatric nurses should educate young people with CF and their parents about self-management of the disease and should support and raise awareness in line with the needs that the family cannot identify during the transition process (Disabato et al., 2019). In order to participate effectively in shared decisions, CF youth need to develop their trust in health care personnel and interact with them. They also need to learn how to manage their condition and treatment on their own as they move into adulthood. Children and young people involved in the joint decision-making process in health services are expected to be more knowledgeable, feel more prepared, and be less worried about the unknown (Malone et al., 2019). The importance of information and preparation for caregivers as well as young people is also emphasized to promote successful transition to adult health care. It is reported that providing parents with clear information and guidance will lead to improvements in transition experiences (Coyne et al., 2018).

Nutritional status has been reported to have a strong positive correlation with lung functions and survival in CF. When growth or nutritional status is impaired, individuals with CF receiving oral nutritional supplements, followed by polymeric enteral tube feeding and complementary enteral tube feeding are recommended to receive continuous night infusion (Schwarzenberg et al., 2016). It is reported that tube feeding leads to weight gain and improves nutritional status and lung functions. Nutrition style, product to be given and the time of administration should be determined according to the preferences of the patient (Hizal, 2019). For the CF team, the main goal of nutrition is to achieve normal growth in children and to maintain adequate nutrition (Castallani et al., 2018). It is the responsibility of pediatric nurses to educate the adolescents with CF and their parents about nutrition in CF (Schwarzenberg et al., 2016).

The health care team should take into account the wishes of the dying patient and their families. Patients may choose to receive hospital care from the staff they know well in a familiar setting. Support at home (e.g. cleaning airways, timely symptom control) is an important consideration to best manage all the symptoms if they want to be at home (Castallani et al., 2018). In the process of preparing the family and the patient for the expected death, their need for

communication, comfort and painless end should be met (Price & Knotts, 2017). From this perspective, it is seen that adolescents with CF and their parents do not have enough information about palliative care. Pediatric nurses need to integrate the educational interventions associated with palliative care into the routine training steps of the CF and fill the gap in performing end-of-life care (Dellon et al., 2018).

As a result, CF requires lifetime nursing care. CF is a chronic disease which has significant effects on children and adolescents and their families. In the management of CF, pediatric nurses should adopt family-centered care, and educate and improve the family to cope with the problems they may encounter during the process.

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