

CASE REPORT

NURSING CARE OF A PATHIENT DIAGNOSED COLD CYSTIC PHYBROSIS ACCORD TO MARJORY GORDON'S MODEL: A CASE REPORT

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ABSTRACT

Cystic fibrosis is a disease that affects many systems as a result of dysfunction of exocrine glands seen in childhood. This disease, which especially affects the respiratory and digestive systems, creates a progressive picture that causes serious problems in quality of life. Therefore, the patient should be handled with a holistic approach. Nurses, who are health professionals, play an important role in the diagnosis and treatment process of the disease. In this case, the patient diagnosed with cystic fibrosis was planned to be nurtured according to Gordon's Health Patterns model. Within the scope of this model, the nursing care of the patient was examined according to 11 functional health patterns. Sick; It has been evaluated in many aspects such as perception, nutrition, excretion, sleep, sexuality and belief. In this case, the purpose of nursing care of a pediatric patient diagnosed with cystic fibrosis; The main purpose of nursing care to be applied to the patient is to relieve the patient's respiratory functions, to provide adequate nutrition, to protect against possible infections, to support growth and development, and to assist the patient and his family in the management of the disease in the process.

Keywords: Cystic fibrosis, nursing care, pediatric patient.

KİSTİK FİBROZİS TANILI ÇOCUK HASTANIN FONKSİYONEL SAĞLIK ÖRÜNTÜLERİ MODELİNE GÖRE HEMŞİRELİK BAKIMI: OLGU SUNUMU

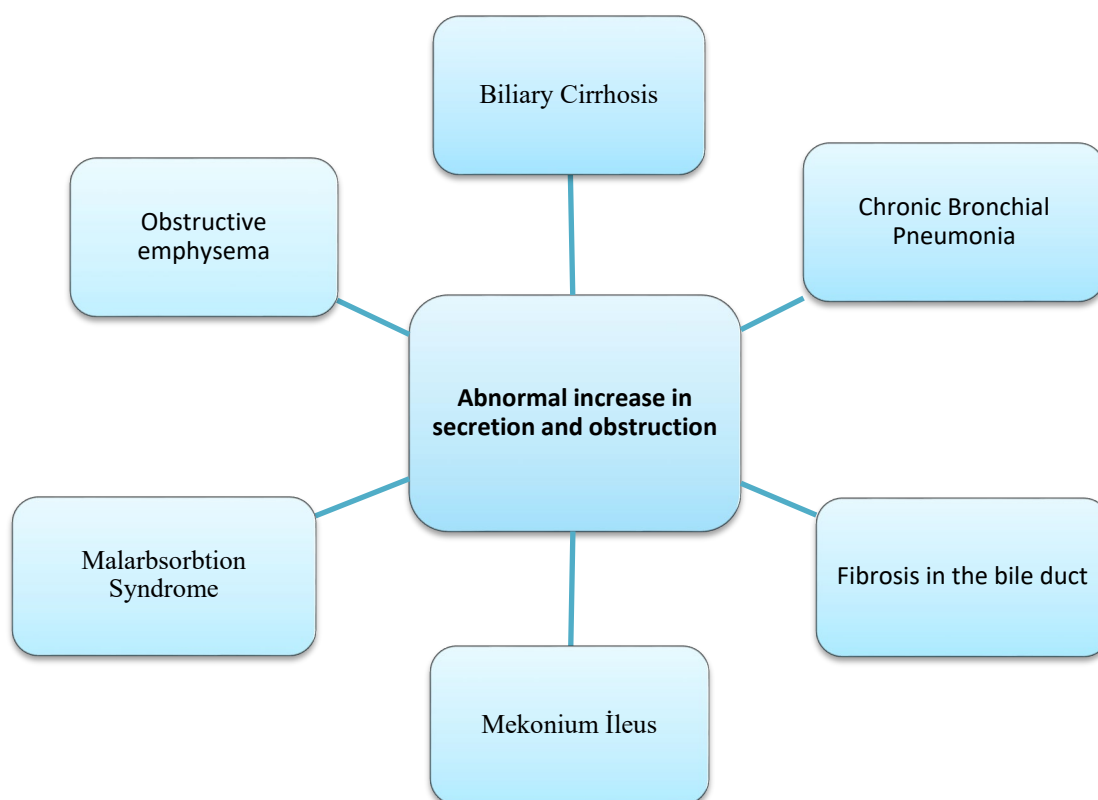
ÖZ

Kistik fibrozis çocukluk çağında görülen ekzokrin salgı bezlerinin disfonksiyonu sonucu birçok sistemi tutan bir hastalıktır. Özellikle solunum ve sindirim sistemlerini tutan bu hastalık ilerleyici ve yaşam kalitesinde ciddi sorunlara sebep olan bir tablo oluşturmaktadır. Bu yüzden hasta bütüncül bir yaklaşım ile ele alınmalıdır. Hastalığın tanı ve tedavi sürecinde sağlık profesyonelleri olan hemşireler önemli rol oynamaktadır. Bu olguda kistik fibrozis tanılı hastanın Gordon'un Sağlık Örüntüleri modeline göre hemşirelik bakımı planlanmıştır. Bu model kapsamında hastanın hemşirelik bakımı 11 fonksiyonel sağlık örüntüsüne göre incelenmiştir. Hasta; algılama, beslenme, boşaltım, uyku, cinsellik, inanç gibi birçok yönden değerlendirmeye alınmıştır. Bu olguda ele aldığımız kistik fibrozis tanılı çocuk hastanın hemşirelik bakımının amacı; hastanın solunum fonksiyonlarını rahatlatmak, yeterli beslenmesini sağlamak, gelişebilecek enfeksiyonlara karşı korumak, büyüme ve gelişmesini desteklemek ve yaşanan süreçte hastaya ve ailesine hastalığın yönetimi konusunda yardımcı olmak hastaya uygulanacak hemşirelik bakımının en temel amacıdır.

Anahtar Kelimeler: Kistik fibrozis, hemşirelik bakımı, pediatrik hasta.

1. INTRODUCTION

Cystic fibrosis is an autosomal recessive disease caused by mutation of genes responsible for coding CFTR protein. There are more than 2000 known gene mutations for the disease (1). The most dominant known gene mutation is F508del. The gene mutation that occurs in cystic fibrosis disease causes a disorder of Cl⁻ transport in organs with epithelial cell structure, primarily in the digestive, respiratory and reproductive systems. As a result, the mucus in the body thickens and prepares the ground for the infection of microorganisms. The most common infections are lung-centered. Condensed mucus causes permanent damage to the lungs (2). 90% of mortality rates are due to lung damage. It has been determined that this disease, which develops once in 2500 births, is more common in the white race than in the black race (2). Turkey was also included in the screening program in 2015 (3). IRT measurement, sweat test, DNA analysis and nasal potential difference measurement are frequently used methods for diagnosis. The IRT value is measured by heel blood. A second IRT measurement is performed for babies with high first measurement results. Afterwards, if the value continues to be high, babies are directed to the sweat test. To collect sweat, pilocarpine is applied to a small area on the arm or leg in children. In the sweat test, electrodes with low electric current are placed in the area to stimulate sweat production. The result of the sweat test was <30 mmol/L in infants under six months of age, and the Cl⁻ value in sweat was accepted as within the normal range. In infants older than 6 months, a Cl⁻ value above >59 mmol/L is considered the diagnostic criterion for cystic fibrosis. The accuracy of the result of this test should also be confirmed by gene analysis (4). Another method used for diagnosis is nasal potential difference measurement. The epithelium of the respiratory tract, including the nasal epithelium, regulates the composition of fluid on the respiratory tract by the transport of sodium and chlorine ions. This transport of ions creates a trans-epithelial difference, and this potential difference can be measured in the nasal epithelium. Ion transport anomalies in the airway epithelium of cystic fibrosis patients create a different nasal potential difference compared to normal epithelium(5). The effects of cystic fibrosis on the body are schematized below (6).



CASE REPORT

The fifteen-year-old male case is the child of a consanguineous family. As a result of the examinations performed in the hospital, where he applied for the first time due to foul-smelling oily stools and retardation in his development, the CI level in sweat was 118 mEg/L. The patient was sent to Istanbul for gene analysis and was diagnosed with cystic fibrosis at the age of 6 months and eight days. Since our case resided in Midyat, after this process, the controls were made at Midyat state hospital. Our patient, who applied to the pediatric outpatient clinic in 2019 due to increasing abdominal pain, vomiting and excessive weight loss, was found to have blood values (aPTT 41.6, INR 1.52, PT 19.4) in the blood tests, and as a result of abdominal USG (liver long axis is 150mm and is larger than normal. The ratio of the liver to the right lobe and left lobe is increased. Liver contours, lobule and parenchyma internal structure (heterogeneous micronodular appearance) with the suspicion of chronic liver disease, he was referred to Dicle training and research hospital, where he was diagnosed with biliary cirrhosis. Our patient, who came to Midyat state hospital to receive 5 mg vitamin k (IV) treatment every week, is followed up as an inpatient due to upper respiratory tract infections once or twice a year. The drugs used by our case are as follows;

Tablo 1. Drug Table

Drug Name	Route Of Delivery	Frequency Of Issuance
Creon 2500 IU	Oral	4*1
Pulmozyme 2.5 mg/2.5 ml	Inhaler	1*1
Polyvit 100 ml	Oral	1*1
Zinco 15 mg /5 ml	Oral	1*1
Dekas plus 60 tb.	Oral	2*1
Ursofalk 250 mg / 5 ml	Oral	2*1
Avikap30 mg	Oral	1*1
K vit. 5 mg	Intravenous	Once a week
Flu vaccine	Intramuscular	Twice a year
PediaSure food 1.5 kcal/ml	Oral	4*1
Evikap 200 IU	Oral	1*1

2.1-) Evaluation of the Child According to Marjory Gordon's Model of Functional Health Patterns

This model, created by Marjory Gordon in 1987, provides the opportunity to consider individuals in biological, psychological and social dimensions. Gordon's Model is a framework used for an individual's health assessments and planning. The model was developed with the aim of ensuring the individualization of health care and guides health professionals in providing specialized care to individuals. Gordon's model includes 11 functional health components. These areas include health status, nutrition, excretion, activity and exercise, sleep and rest, cognitive and perceptual status,

self-perception, roles and relationships, sexuality and reproduction, ability to cope with stress, and patterns of values and beliefs.

2.1.1-) Health Perception - Health Management

Our 15-year-old patient does not have enough information about the diseases. Our case, who carried out the treatment process as dependent on his family, stated that he had a lot of trouble due to hospitalizations, and that he sometimes delayed in telling his family about the complaints he felt about his illness because he did not like the hospital environment. Her parents stated that they did not know how to proceed with the progress of the treatment, so they carried out the treatment as told by the doctors, and that they were afraid because they lost one of their children due to cystic fibrosis.

Nursing diagnosis 1: Lack of Knowledge

Area 5: Sensing / Grasping

Class 4: Grip

Descriptive Features: The family expressing that they do not know how to proceed with the treatment progress, expressing their complaints about the child's illness late.

Objective: To eliminate the lack of knowledge of parents and children about the disease and the treatment process.

Initiatives:

- Issues with lack of information are identified.
- It is ensured that the family and the child express themselves comfortably and relieve their concerns.
- Uncertainties related to the disease and the treatment process are identified.
- Communication is established with the child about the importance of the child voicing his complaints.

Evaluation: The knowledge of the family and the child about the complications of the disease and the treatment process was evaluated as 4 out of 5 after the trainings.

Nursing diagnosis 2: Fear

Area 9: Coping / Stress tolerance

Class 2: Coping

Descriptive Features: Expressing fear for the loss of the family due to this disease.

Purpose: To enable the family to express their fears freely.

Initiatives:

- It is ensured that the family expresses their feelings about the loss they have experienced.
- Information is exchanged with the family about the reasons for the fears they experience.
- If there are fears that these children experience due to the unknown about the treatment process, the unknowns about the treatment process of their children are eliminated.

Evaluation: Psychological relief was observed in the family as they comfortably expressed their fears to a health professional. It was determined that the fear of death for their children still continued.

2.1.2-) Nutrition and Metabolism

Our patient, who often experienced nausea and burning sensation in the stomach, stated that he did not like to eat very much. 44 pounds (5.ve 10. The parents of our patient described their child

as having no appetite and stated that they drank 4 pieces of pediasure formula a day and thus their child was better.

Nursing diagnosis 3: Nutrition Less Than Body Requirement

Area 2: Nutrition

Class 1: Food intake

Descriptive Features: Expressing that he does not like to eat because of nausea and a burning sensation in the stomach. It is between the 5th and 10th percentile values.

Objective: To ensure that he has the ideal body weight. Collaborating with a dietitian for adequate calorie intake.

Initiatives:

- Body weight is monitored.
- The dishes he likes are questioned and the meal order is planned accordingly.
- By cooperating with a dietitian, it is ensured that our case reaches the ideal weight.

Evaluation: As a result of the cooperation with the hospital dietician, the weight of our patient increased from 40 kg to 44 kg in a period of two and a half months. The ideal body weight has not yet been reached.

Nursing Diagnosis 4: Nausea

Area 12: Comfort

Class 1: Physical comfort

Descriptive Features: Feeling nauseous while eating, problems with the bile duct.

Objective: To determine the causes of nausea, to relieve nausea.

Initiatives:

- It is ensured that foods with high spice and fat content are avoided.
- It is recommended to feed at small and frequent intervals.
- After eating, they are informed not to lie flat for 2 hours.
- It is said that fluids should be restricted with food.
- If vomiting occurs, the number of times he vomits is recorded and the doctor cooperates. If necessary, antiemetics are applied at the request of the doctor.

Evaluation: It was observed that he did not consume liquids while eating. As a result of staying away from spicy and fatty foods, it was evaluated by his parents that his nausea, which he experienced frequently daily, decreased.

2.1.3-) Excretion

Our case, who stated that he did not have any problems while urinating, stated that he sometimes felt pain while pooping and had difficulty in doing so. His family also stated that he often had constipation problems.

Nursing Diagnosis 5: Constipation

Area 3: Discharge and Gas exchange

Class 2: Gastrointestinal Function

Descriptive Features: Expressing that he has constipation problems, having difficult stool output.

Objective: To ensure that they gain regular bowel habits.

Initiatives:

- Risk factors for constipation in the case are determined.
- The intake of fibrous foods is increased.
- Fluid intake is increased.
- The importance of not postponing defecation is explained to the patient.
- If necessary, laxatives are applied at the request of the doctor.
- It tells the importance of defecating at the same time every day.
- The importance of adequate exercise is explained, walking is recommended.
- The importance of avoiding psychological problems such as stress and anxiety is explained to the child and their parents. (Stress is an important factor in constipation.)

Evaluation: Pelvic floor muscle exercises were performed on the patient for two months. The proportion of vegetables and fruits in food intake was increased. Daily fluid intake (fluid intake of less than 500 ml was minimized, 1000 ml was increased). It was observed that the patient's defecation output was more relaxed compared to the previous months.

2.1.4-) Activity and Exercise

Our case, who reads openly due to his illness, spends most of his time on the phone. Our case said that he sometimes went to play football with his friends. His family stated that he was very sick with respiratory diseases, so they had breathing exercises, but this was not done regularly.

2.1.5-) Sleep and Rest

The family of our patient who had difficulty in expectorating sputum stated that they had difficulty in this regard for both themselves and their children at night, especially after steaming (postural drainage). Our case, who said that he liked to sleep during the day, described himself as constantly sluggish and tired.

Nursing Diagnosis 6: Disruption in Sleep Pattern**Area 4: Activity/Recreation****Class 1: Sleep-Rest**

Descriptive Features: Expressing that his/her sleep is interrupted due to drug intake.

Objective: To ensure the development of a rested sleep routine.

Initiatives:

- Drug treatments are taken to the times when the child is awake with the supervision of the doctor.
- It is recommended that postural drainage be performed in the left posterior segment of the inhaler to be given at night.
- It is tried to gain the habit of activity that will increase sleep quality.
- A relaxing, quiet and calm environment is created.
- An activity program is prepared to keep him awake during the day, and daytime sleep is shortened.

Evaluation: The daytime sleep of our daytime sleeper was reduced from approximately two to three hours to one to one and a half hours. The frequency of playing with the phone while going to bed at night has been reduced. Parents were trained in postural drainage in the left posterior segment. Our case rated the current sleep quality as 3 out of 5.

Nursing diagnosis 7: Fatigue

Area 4: Activity/Recreation**Class 3:** Energy Balance

Descriptive Features: Our patient feels tired and sluggish, sleep disorder occurs.

Initiatives:

- If the individual has fatigue due to malnutrition, this situation is reported to the dietitian and physician and a protein-based diet is prepared accordingly.
- The factors that cause fatigue are investigated.
- Blood-hormone levels are checked for the patient's symptoms of weakness.

Evaluation: Vitamin D was found to be 20 in the blood test and vitamin D treatment was started. By changing the sedentary lifestyle, the patient felt more vigorous and energetic (enrolled in the gym).

2.1.6-) Cognitive Perception

Our case has no problem in place, time and person orientation. Our patient, who did not have problems in taste, smell, touch and hearing, said that he had problems in the eye. According to his family, he takes medication once or twice a year for conjunctivitis. Our patient, who does not have any mental problems, has cognitive and affective skills brought by his age. Our phenomenon, which has a timid facial expression, does not have a problem in expressing itself.

Nursing Diagnosis 8: Risk of Infection**Area 11:** Security / Protection**Class 1:** Infection

Descriptive Features: Cystic fibrosis, conjunctivitis and hospitalization for respiratory tract infections two to three times a year.

Objective: To protect the patient against existing infection risk factors and to provide information on this subject.

Initiatives:

- Hygiene training is given to the family and the patient to prevent the development of infection.
- The patient's blood tests (crp, wbc) are followed.
- Contact with sick people is cut off to prevent respiratory infections.
- It is said that the patient should not share personal care items with anyone else and should not use someone else's personal belongings.

Evaluation: No infection was observed in the patient.

2.1.7-) Self-Perception and Self

Our fifteen-year-old patient said that he went to the hospital all the time and felt both tired and angry because of this situation. Most of them stated that the decision was not consulted with them, that they always lived the way their family or doctors told them, that they did not feel free, that they were stressed. This is why our case in adolescence has expressed that he gets angry quickly and his parents have stated that their child immediately becomes aggressive.

Nursing diagnosis 9: Stress Overload**Area 9:** Coping / Stress tolerance**Class 2:** Coping

Descriptive Features: Development of anger problems as a result of stress, being in adolescence.

Objective: To identify the stress factors that cause anger and to counsel the patient.

Initiatives:

- Participation in the own treatment process is ensured.
- The stressors that cause him to get angry are identified.
- Decisions that will concern his own life are made with his participation.
- Parents are informed about the adolescent period.
- Psychological support is provided for him to express himself comfortably.

Evaluation: As a result of the adolescent education given to the family, a difference was observed in the family's approach to the child. He actively participated in the decisions taken during the treatment process.

2.1.8-) Role-Relationship

Our case, who said that he continued his high school education openly because he had to go to the hospital frequently, stated that he liked to go to school and study formally, and that he missed his friends.

Nursing Diagnosis 10: Ineffective Role Performance

Field 7: Role relationships

Class 3: Role performance

Descriptive Features: Attending school openly due to hospital controls.

Objective: To provide support in performing their roles and performances.

Initiatives:

- It is ensured that he expresses himself comfortably.
- The factors that cause ineffectiveness in the role performance of the individual are investigated.
- They are encouraged to talk about their friends and relationships.
- Even if he cannot read formally, his friends and his participation in social activities are discussed with the family.
- They are supported to attend courses in the fields they love.

Evaluation: Our case, who is interested in photography, attended the photography course with referrals. It was observed by their parents that it positively affected their roles and relationships.

2.1.9-) Sexuality and Reproduction

Our case, who was fifteen years old, was in adolescence. He said that he did not have a disorder in the reproductive system and expressed his interest in bilateral relations. She has stated that she has had some flirtation periods. Pupik hair growth (+) and thickening of the voice (+) specific to the period in which it is located.

10-) Coping – Stress Tolerance

He said that he normally has a calm nature, but sometimes he is angry about the hospital. He stated that when he was stressed, he listened to music and played games on his phone, so he relaxed.

11-) Value- Belief

His parents said that they occasionally sent their children to religious education courses. Our case stated that he liked to pray.

3.DISCUSSION

Cystic fibrosis is an autosomal recessive disease with a carrier frequency of 1/25 and a live birth rate between 1/2000-1/3500 (7). The incidence of this disease in Turkey is unknown. The opinion as a result of the researches is that it is a rare disease in our country. The incidence of this disease, which is autosomal recessive, increases with consanguineous marriage (8). According to the 2022 consanguineous marriage data shared by TurkStat, the rate of consanguineous marriage in our country decreased from 5.9% to 3.9% (9).

Since cystic fibrosis affects many systems such as respiratory, digestive and reproductive, health counseling should be provided to these patients and the people responsible for their care. For this, nurses play an active role.

In this case, the nursing care plan of a 15-year-old adolescent child was created according to Marjory Gordon's functional health patterns model. Gordon's model addresses 11 health functions and thus makes it easier for nurses to address the patient physically, psychologically, and socially. This model actually enables nurses to evaluate the patient from a holistic perspective and plan nursing care for him.

The important thing in nursing care is to guide parents about the issues to be considered in the care of the child. According to a study conducted in 2015, the destructive effect of the disease, especially on the respiratory and digestive system, will produce positive results with good care and treatment, and the quality of life and average life expectancy will increase (10).

In this case, parents were informed about breathing exercises and postural drainage in order to gain a better function of respiratory functions. It is aimed to further relieve breathing and prevent condensed mucus from developing an infection in the lung. In a study conducted in 2003, it was determined that the mortality risk of cystic fibrosis patients was due to lung involvement (11). The family was informed about the importance of chest physiotherapy after inhalers.

In a study conducted in 2021, cystic fibrosis, malabsorption and digestive system problems in patients cause malnutrition and reduce the body's muscle volume ratio. Due to this situation, the contraction forces of the muscles are affected, in which case it negatively affects the lung and immune system. According to another study conducted in 2016, gastrointestinal system problems are seen in 85-90% of patients with cystic fibrosis (12). For this reason, nutrition is very important for patients with cystic fibrosis. Providing nutrition counseling to parents is carried out in cooperation with nurses, doctors and dieticians. For our patient who had nausea and constipation problems, constipation and nausea diagnoses were discussed in the nursing care plan in cooperation with the doctor and dietician. For our case, which was in the 5th percentile in terms of body weight, a diet program with high energy content was created with the cooperation of a dietitian

Since our case, which was evaluated psychologically and socially, was in the adolescent period, it sought to be independent from the parent specific to this period, and it was observed that the state of dependence in terms of the treatment process triggered the state of anger and this situation wore out the caregivers. It is very important for nurses to support the patient and parents emotionally at these times and to inform the family about the characteristics of the adolescent period.

Studies have shown that the approach that will make it easier for caregivers to cope with distressing processes for children with chronic diseases such as cystic fibrosis is the parent-centered care approach. Parent-centered care enables parents and nurses to progress more effectively and in a planned manner based on the cooperation of child care services. Parent-centered care approach, which is one of the basic principles of pediatric nursing. The Association for Care of Children's Health has

defined it as a philosophy of care that puts the family at the center of life throughout the child's illness and recovery process (13).

4. CONCLUSION AND RECOMMENDATIONS

Cystic fibrosis, which is thought to increase with the increase in consanguineous marriage, is a chronic disease in which the quality of life and average life expectancy can be increased thanks to a holistic nursing care that will include the parent and the child. In this case, nursing care was handled in terms of 11 health functions and the negative effects of this disease on the child and the parent were minimized. The positive effects of nursing care offered from a holistic perspective on children with chronic diseases and their parents were observed in this case, and nursing care was concluded positively with the active participation of our patient and their parents.

Declaration of conflicting interests: None.

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