

Case Report

Nursing Care of a Child with MOPD II and Moyamoya according to Orem's Self-care Deficiency Theory: A Case Report

Orem'in Özbakım Yetersizliği Teorisine Göre MOPD II ve Moyamoya'lı Bir Çocuğun Hemşirelik Bakımı: Olgu Sunumu

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ABSTRACT

Microcephalic osteodysplastic primitive dwarfism (MOPDII) is the most familiar form of microcephalic primordial dwarfism (MPD). The classic features of the MPD group are marked microcephaly and severe antenatal and postnatal growth retardation. Such patients also have Moyamoya disease, which includes recurrent stroke attacks and possible neurological complications. Today, it is possible to provide care for such patients with many theories. Caring for children with growth-developmental retardation with Orem's Self-care Deficiency Theory may enable them to become self-aware and increase their quality of life. The significance of nursing care for this rare disease is undeniable. In this study, the nursing care of a patient diagnosed with MOPD2 and Moyamoya was carried out based on the Self-care Deficit Nursing Theory. "Partly remedial nursing care" was provided as the patient can self-care at a moderate level, and also "supportive and educational nursing care" was provided to support the mother and other family members who provide care to the patient.

Keywords: *Moyamoya, Microcephalic Primordial Dwarfism, Child, Nurse, Orem's Self-care Deficiency Theory*

ÖZET

Mikrosefalik osteodisplastik ilkel cücelik (MOPDII), mikrosefalik primordial cüceliğin (MPD) en bilinen şeklidir. MPD grubunun klasik özellikleri, belirgin mikrosefali ile ciddi doğum öncesi ve sonrası büyüme geriliğidir. Bu tür hastalarda olası nörolojik komplikasyonları vurgulayan tekrarlayan inme ataklarıyla karmaşık hale gelen Moyamoya hastalığı eşlik eder. Günümüzde birçok teori ile bu tür hastalara bakım sağlamak mümkündür. Büyüme- gelişme geriliği olan bu çocuklarda Orem'in Öz Bakım Eksikliği Kuramı ile bakım vermek çocuğun kendinin farkına varmasını ve yaşam kalitesinin artmasını sağlayacaktır. Nadir görülen bu hastalıkta hemşirelik bakımının önemi yadsınamayacak düzeydedir. Öz-Bakım Eksikliği Hemşirelik Kuramı'nı temel alarak MOPD2 ve Moyamoya tanısına sahip hastanın hemşirelik bakımının yürütüldüğü bu çalışmada hasta öz-bakımını orta düzeyde yapabildiği için "kısmen eksikliği giderici hemşirelik bakımı" ve hastanın öz-bakımını karşılayan annesini ve diğer aile üyelerini desteklemek için de "destekleyici ve eğitsel hemşirelik bakımı" uygulanarak bakım sağlanmıştır.

Keywords: *Moyamoya, Mikrosefalik Primordial Cücelik, Çocuk, Hemşire, Orem'in Öz Bakım Eksikliği Teorisi*

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INTRODUCTION

Microcephalic osteodysplastic primitive dwarfism (MOPD II) is the most common and best-described form of microcephalic primordial dwarfism (MPC). The classic features of the MPC group are marked microcephaly and severe antenatal and postnatal growth retardation (1-4).

Children with MOPD II are dependent on others throughout their lives due to growth and development delays, and this negatively affects their ability to self-care and their quality of life. In this study, due to its rarity in Turkey, the case of MOPD II was addressed. Nursing care was planned using Orem's Self-care Deficiency Theory since children with this diagnosis cannot self-care and always need a caregiver. This study aimed to provide better care to children diagnosed with MOPD II and Moyamoya, to present an example of a nursing care plan based on Orem's Self-care Deficit Theory so that they can cope with the disease, and thus to guide and provide consultancy to the nurses.

CASE REPORT

Key Situational Factors

1. Pregnancy and birth history

S.A. is born at 35 weeks+6 days with normal birth. She was 35 cm tall (<3rd percentile) and weighed 920 g (<3rd percentile). S.A. remained in the incubator for 36 days. She was discharged at 1150 g.

2. Disease history

The baby's weight gain and height growth were very slow until the sixth month. After the sixth month, she was diagnosed with MOPD II and Moyamoya, and the treatment process was started (Carbamazepine 3x1, Levothyroxine sodium 1x1, Acetylsalicylic acid 1x1). At 18 months, S.A. had a seizure, developed partial paralysis in her left hand and foot, and recovered after two weeks of physical therapy. She started attending special education when she was 22 months old. She has been continuing her education for eight years.

Current patient information:

Age: 13

Gender: girl

Diagnosis: MOPD II, Moyamoya

Weight: 24.5 kg (<3p) **Height:** 81cm (<3p) **Head Circumference:** 43 (microcephalic appearance)

Socio-cultural characteristics: SA has a nuclear family and has two siblings. Siblings of SA have no known disease.

Lifestyle: She can walk but gets tired quickly due to motor retardation. Although she can do many things herself, she gets support from her family because she feels exhausted. She goes from one place to another on her mother's lap or with a baby carriage. She does not like to eat, especially because she has problems chewing and swallowing food.

Interaction with the environment: Social isolation has increased during the pandemic period. He continues private education at home. There is physical and motor skill retardation.

Presence and adequacy of resources: She has social security. Their economic status is moderate.

4. Self-care

Since the child cannot fully perform self-care activities independently, she is partially dependent on the family. However, the child's self-care agent is primarily the mother.

5. Self-care strength

The level of meeting the child's self-care needs is low. Needs help in meeting self-care needs.

Self-confidence and respect: The child established eye contact while speaking. She is extrovert and sociable.

Ability to control and initiate her energy: She can perform activities of daily living at a moderate level. She has difficulty breathing when she does long exercises and walks.

Comprehension competence: Perceptual and cognitive status is moderate. The child generally understands what is being said and can do what is asked. She has difficulty making sense of some newly heard concepts. She can become irritable and stubborn when she does not understand. She is easily distracted.

Motivation: The child can express her wishes and problems verbally. When she is stubborn and cannot make sense of some things, the mother tries to motivate her by repeating or explaining the same information over and over again.

Making decisions about self-care: The child can perform basic self-care skills with help, and the mother

usually makes the decisions about the child, except for the choice of clothes to wear.

Perceptual, cognitive status and communication skills: The child gives the desired answers to the questions. She plays cooperative games with the other children. She has trouble understanding new concepts. She fulfills her responsibilities in a game properly. She communicates well with her family as well as her environment.

6. Therapeutic self-care needs

Therapeutic self-care needs are examined under three sub-dimensions.

I. Universal self-care needs

Air: She has difficulty breathing because her lungs are not fully developed. The respiratory distress of the child increases, especially with excessive movement or exercise.

Nutrition: She can eat by herself. She mostly enjoys consuming foods that she can swallow without chewing (e.g., soup, pudding and yogurt).

Oral health: Her teeth are sparse and small.

Activity: The child can walk short but has difficulty climbing stairs. She can have her bath with help. Her physical activity is moderate.

Sleep: Total sleep time at night varies between 7-8 hours. She has difficulty falling asleep.

Cognitive perception: The child's level of perception and reaction to events is medium.

II-Developmental self-care needs: The child has a physical and motor developmental delay. He does not like crowds. The pandemic has increased the social isolation of children and families.

III- Health deviation self-care needs:

- Difficulty in breathing
- Disruption in sleep pattern
- Vision problem
- Hearing problem
- Nutrition less than body requirement
- Disproportionate growth
- Oral motor problems
- Social isolation

7. Lack of self-care

The power of the dependent care agent: Communication between child and mother is better than other family members. The mother is knowledgeable about her child's illness and needs.

Self-care deficit of the dependent care agent: Due to the mother's other responsibilities, she has difficulty allocating enough time to SA.

8. Nursing Process

In our study was determined that the child was deficient in self-care skills. Thus, "Partly Remedial Nursing Care" was given to the child and "Supportive and Educational Nursing Care" was given to his family (5-14). Informed onsent was obtained to conduct this case study.

Nursing Diagnosis	Aim	Nursing Intersentions	Evaluation
Disproportionate growth due to height and weight below the 3rd percentile and inability to perform age-appropriate behaviors and skills (NA ND A)	To monitor and support growth and development in line with the goals set by the Treatment and Care team	<ul style="list-style-type: none"> • The importance of providing an environment and opportunities that will enable the child to perform developmental tasks appropriate for her age period was explained to the mother. • It was stated that the child should be supported to perform self-care skills independently. • It was stated that rewarding the child's positive behaviors would have an impact on her development. • The importance of regular physical and muscle strengthening exercises recommended by the physiotherapist in order to increase her physical development was explained 	<ul style="list-style-type: none"> • It was observed that she regularly performed the recommended exercises and muscle strengthening exercises recommended by the physiotherapist. • Since the maintenance period was short, sufficient change in growth could not be observed.

Nursing Diagnosis	Aim	Nursing Interventions	Evaluation
Disruption in sleep pattern	To make it easier for the child to fall asleep and to improve the quality of sleep.	<ul style="list-style-type: none"> • Factors affecting the child's sleep were identified. • It was explained that creating a quiet, calm and relaxing sleep environment would increase the quality of sleep. • Interventions were planned to make it easier for her to fall asleep. • The importance of proper sleeping position and comfortable breathing was explained. • It was explained that drinking warm milk before going to sleep, listening to soothing music or reading a book would make it easier to fall asleep. 	<ul style="list-style-type: none"> • The mother stated that she started to give S.A. warm milk before going to sleep at night. • S.A.'s mother or older sister started reading a story book before she went to sleep. • SA stated that she had no difficulty falling asleep. • There was no change in sleep time. She sleeps for an average of seven hours
difficulty in breathing	To ensure easy breathing	<ul style="list-style-type: none"> • An age-appropriate explanation was made about the importance of deep breathing exercises. • It was explained that saline can be applied to prevent nasal congestion. 	<ul style="list-style-type: none"> • The child said she would do the deep breathing exercises.
Nutrition less than body requirement due to difficulty in chewing and swallowing food (BMI <3 percentile)	To ensure that the child is fed in accordance with her daily metabolic needs and activity level.	<ul style="list-style-type: none"> • She was told that she should receive support from a dietitian in order to have an adequate and balanced diet suitable for her age. • It was stated that the foods that she likes should be added to the diet list and the foods that she dislikes should be minimized • It was explained that using therapeutic play techniques while eating would have positive effects on the child's food consumption. The therapeutic play techniques were explained to the mother (storytelling, creative play etc.). • It was stated that it would be effective to use methods such as competition and rewarding while eating with siblings. 	<ul style="list-style-type: none"> • A meeting with a dietitian was arranged for SA. in the rehabilitation center she attends regularly. • The mother stated that giving food to SA. with stones worked and SA. was happy, but she stopped eating when she was tired • SA. refused to compete with her siblings because they eat very quickly
Lack of self-care due to the inability to perform activities of daily living	To ensure the highest level of participation of the child in self-care practices	<ul style="list-style-type: none"> • It was stated that rewarding for participating in care is important in reinforcing this behavior. • Daily routines were created for activities such as eating, brushing teeth, and getting dressed, and daily care follow-up was made thanks to the prepared lists. • It was ensured that she could develop her skills by choosing the appropriate clothes and by always wearing and removing (the same clothes so that she could get dressed on her own 	<ul style="list-style-type: none"> • It was observed that SA. started to participate more actively in her daily self-care in line with the lists prepared • Her participation in each self-care activity was evaluated and she was rewarded for completing the 4-week care list completely. • The development of fine motor skills was supported by using specially prepared materials for the development of self-care skills such as wearing shoes, buttoning and tying laces.

Nursing Diagnosis	Aim	Nursing Interventions	Evaluation
Change in families as a result of having a child with a chronic health problem	To create a functional system where family members support each other.	<ul style="list-style-type: none"> • The mother, father and siblings were supported to understand the needs of the sick child and to make necessary arrangements, and the family was given training on how to manage the process more effectively. • The family was informed about the need to avoid over-protecting the child and the importance of supporting the child so that she can do the things she can. • The mother was encouraged to take time for herself while S.A. was at the rehabilitation center. • The importance of doing activities where all members of the family will spend time together was explained. • It was explained that it would be beneficial for them to meet face-to-face or over the phone with other families with similar problems. 	<ul style="list-style-type: none"> • To reduce the burden of the mother, who is the primary care giver, the father and the older sister began to help S.A. cat in the evenings. • Every evening, family members take turns reading a book to S.A. • The mother stated that her burden eased a little when she shared the care of S.A. with other family members.
Hearing problem	To ensure that the child expresses that she hears better	<ul style="list-style-type: none"> • The importance of using hearing aids to prevent possible traumas was explained to the child and her family. • The importance of keeping the room of the child with hearing problem bright and having a night light on to illuminate the room at night was explained. • It was explained that it is necessary to stand in a suitable position where eye contact can be established with the child during communication. • It was stated that the surrounding noises and distractions should be eliminated before communicating with the child. 	<ul style="list-style-type: none"> • The child and her family was informed about the use of hearing aids. • S.A. stated that she had difficulty in wearing the device because his auricle was small. • S.A.'s mother said that they used a night lamp in her room. • It was observed that the mother was at eye level while talking to the child.

DISCUSSION

MOPD II and Moyamoya require a multidisciplinary approach, especially for nurses with significant responsibilities. Although many studies are related to treating children diagnosed with MOPD II, to our knowledge, no studies have been found addressing nursing theories and care (5-8). Providing care using nursing theories is crucial in providing a comprehensive, systematic approach and a holistic nursing perspective at every stage of care (9-14). It ensures that patients can be cared for in an evidence-based and professional manner (5-9).

In the case under consideration, it was observed that the child was able to perform activities of daily living at a moderate level, was inadequate even in basic self-care skills, such as getting dressed, and having a bath, and had nutritional deficiencies and sleep problems. For the child to perform self-care activities independently, responsibilities appropriate to her physical capacity were given in cooperation with his family. Due to the training provided to the child and the family members, the child could put on and take off his clothes and tie shoelaces. S.A., who had difficulty falling asleep, did as she was told, yet there was no change in her sleep duration. The social isolation of the child,

who has been away from the rehabilitation center for a long time due to the pandemic, continues. Thanks to the planning made with the family, it was observed that other family members reduced the mother's responsibilities and burden, and the mother created time for her self-care. When the case was evaluated in general, it was observed that the child started to meet her self-care needs. It was observed that the quality of life of both the caregivers and the child has increased. Orem's Self-care Deficiency Theory provides a general framework directly related to nursing functions and guides nurses. It can be recommended to have a more extended training period to evaluate the behavior and development of the child more comprehensively by using Orem's theory.

Patient Consent Form / Hasta Onam Formu

The parents' of this patient consent was obtained for this study.

Conflict of Interest / Çıkar Çatışması

The authors declared no conflicts of interest with respect to authorship and/or publication of the article.

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