



Communication with acute intermittent porphyria patients who have tracheostomy

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Abstract

Objective: Introducing acute intermittent porphyria disease and investigating the communication techniques for acute intermittent porphyria patients who have tracheostomy

Case Report: 26 years old woman patient consulted to the hospital when she was 13 years old with nausea and vomiting complaints. In 2008, because of respiratory arrest, tracheostomy was implemented to the patient who had acute intermittent porphyria diagnose. The patient had two bullous lesions on the left hand and hyperpigmentation's on the face and legs. As a result of metabolic acidosis (because of urine disease), the patient was taken to dialysis. After six months of dialysis, hypernatremia was found in the laboratory tests (Na: 85). Period of acute crisis developed in the patient with hypertension, atrophy in the muscles, fainting, abdominal pain, nausea, vomiting and diarrhea. Because of serious diarrhea, TPN was started for the patient who was fed with gavage before. Laboratory test results are Na:126, K:8.1, BUN:23.83, Urine:51, Creatinine:0.94, Leukocyte:20.4, erythrocyte:2.22, Hb:6.87g/dl, Hct: %20.9, Plt:106.

Result: It is really difficult to communicate with these patients with lifespan limited because of an extremely severe clinical condition and tracheostomy.

Keywords: acute intermittent porphyria, tracheostomy, communication

Introduction

AIP occurs because of partial defect in the activity of porphobilinogen (PBG). As a result of patient's taking some drugs (sulpha drugs, syrups with alcohol), surgery or anesthesia, patients life could get in danger (Whatley et al, 2009). AIP passes with autosomal dominant inheritance.

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Genetical defect is on the 11th chromosome which describes PBG deaminase. %90 of AIP patients live biochemically and clinically normal for whole their life. However, facts like hunger, alcohol, enfection, drugs like barbiturate, sulpha drugs, hormonal changes especially seen in the postpubertal women can cause the clinical symptoms. During the acute attack, stomach ache, nausea, vomitting, diarrhea and as a result of these, dehydration can occure (Peters & Sarkany, 2005; Bylesjo et al, 2009). We can see tachycardia and hypertension in the %70, tension and cramping pains because muscle weakness in the %70 of these patients (Bylesjo et al, 2009; Periasamy, 2002). During the acute attack, anxiety, depression, psychosis can occure. In some cases, the hypertension during the AIP attack can be the only symptom and the other symptoms can occure later (Aggarwal, 1994; Özer, 2004). Even hypertension can be the first finding of chronic renal failure caused by AIP. In the treatment, high-calorie diet is recommended to stop the catabolic process and iron supplements (panhematin 4mg/kg once in 12 hours or normosang 2-3 mg/kg once in 6 hours) are recommended to give the final product that can't be made (Periasamy, 2002; Peters & Sarkany, 2005). The progression of the disease is very hard and slow after the diagnose of AIP. Furthermore, AIP is a very rare seen disease. The incidence of AIP is 1/10.000 in Sweden and 1/50.000 in the other ethnic groups Bylesjo et al, 2009.

Case Report

26 years old woman patient consulted to the hospital when she was 13 years old with nausea and vomiting complaints. For 1-2 years her disease was investigated, and when her complaints got clear, AIP has become the exact diagnose her treatment started. In 2008 because of respiratory arrest, tracheostomy was implanted and regular follow-ups continued. The patient consulted to the Duzce University Hospital for urinary tract enfection in 2009. After the hospitalization in the urology service for 2 weeks, the patient was moved to the internal intensive care unit for respiratory distress and deterioration of general condition. Period of acute crisis developed In the patient with hypertension, atrophy in the muscles, fainting, abdominal pain, nausea, vomiting and diarrhea. The patient had two bullous lesions on the left hand and hyperpigmentation's on the face and legs. As a result of metabolic acidosis(because of urine disease), the patient was taken to dialysis. After six months of dialysis, hyponatremia was found in the laboratory tests(Na:85). Laboratory test results are Na:126, K:8.1mg, BUN:23.83mg/dl, Urine:51, Creatinine:0.94, Leukocyte:20.4mm³, erythrocyte:2.22mm³, Hb:6.87g/dl, Hct:%20.9, Plt:106. In order to regulate the patient's hypertension(180/100mm/hg), Norvasc (Amladipine) 1x1 was used. Also, for vitamin supplements, Bemix (multivitamin) and Cernevit (multivitamin) flk 1x1, for hyponatermia, Kalinor

(potassium citrate, potassium carbonate) 1x1 and for anxiety, Porozac (Fluoxetine) liquid 1x1 was used.

As a result of the extremely severe clinical condition and the tracheostomy of the patient in the case report, anxiety caused the patient to express herself difficultly and that's why accordance to the treatment was affected badly. Communication is extremely difficult with these patients whose life get in danger in the acute crise periods and whose disease goes with extremely serious symptoms. Because of the tracheostomy, being in the intensive care unit, increased anxiety and severe clinical situation, communication with the patient was extremely difficult.

Discussion

Generally in the intensive care unit, verbal and nonverbal(confirmation, writing, communication cards and touching) communication techniques are used for patients with tracheostomy. During the right communication technique selection, cultural structure, perceptivity, language, age, education level and communication skills of the patient are important. Different techniques can be used in the same patient (Alasad & Ahmad, 2005). More than one communication technique are used in the case. First, marking and confirmation techniques were used as the patient could understand the words but couldn't use the verbal techniques. But, impenetrability and according to this, prolongation of the communication process developed. Communication with marking and confirmation is carried by with nurse's questions and the patient's approvals. For example if the nurse asks the patient as "do you have pain?", the patient answers the nurse "yes" or "no" with eye movements. This technique can be used easily when the other ones can not be used or the nurse can understand the patient easily and correctly (Hemsley 2001; Yava, 2006). But in this technique, the communication should go to the way that the nurse wants according to the experiences, not the way that the patient wants. Because of these stressful and prolonged situations, writing or if necessary training is given communication cards can be used if it is needed (Hemsley 2001;Yava, 2006). In the case, communications cards were decided to be used as the patient couldn't fully express the requests.

The communication cards are very useful in especially patients who don't know the language or who have hearing disability and also the communication cards can be used in all the patients with tracheostomy. These cards have different pictures representing feelings and needs. The patient chooses the card which represent the right feeling and shows it to the nurse. The disadvantages of this technique are that the cards can represent limited feelings or needs, some cards are difficult to understand and preliminary study with the patient is needed to use these cards (MNThóra & Hafsteindóttir, 1996; Yava, 2006). In the case, the cards got ready to communicate with the patient.

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The first subject the patient tried to tell with the cards was "I am very bored. Can you read me a book?". The patient was read a book. It was recognized that the patient listened to the nurse very carefully while readied the book. But as the time passed, she expressed with the cards that she was bored with the book reading and wanted to watch television. It could be seen that she could express her feeling and needs with the cards more easily and the communication was kept on with the cards.

Result

Communication with the patient supports the increasement of the quality of care and satisfaction and also patient's compliance to the treatment.

In this case, it was understood that the right communication technique makes the communication much easier. Not only has the right selection of the communication technique for the patient's characteristics but also the preliminary information and study made the communication easier.

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