

1015

THE CRITICAL THINKING DISPOSITIONS AND EFFECTING FACTORS OF THE NURSES WORKING AT PEDIATRIC UNITS

N. Cinar¹, Ö. Dogu², S. Kuguoglu³

¹Sakarya University, School of Health Sciences,

²Sakarya Education and Research Hospital, Sakarya, Turkey, ³Acibadem University, Health Science Faculty, Part Time Faculty, Sakarya, Turkey

Objective: This study is carried out in order to determine the critical thinking dispositions of the nurses working at the pediatric units and to determine the factors effecting these levels.

Method: The sample of this research is built up by the 36 nurses working for pediatric units in a State Hospital in Sakarya / Turkey (2010). Data were collected using a personel information form and California Critical Thinking Disposition Inventory (CCTDI). In the analysis of the data mean, percentage, Mann Whitney U Test and Kruskal wallis Test were used.

Results: It is determined that 38.9% of the respondent nurses was from neonatal intensive care unit, 36.1% was from general pediatric units, 47.2% was worked 1-5 years in pediatric unit. The average critical thinking disposition score of the participants was found 253.03 ± 19.86 . When the subscale scores were evaluated, the nurses were received the highest mean scores from the analyticity subscale (48.33 ± 4.99) and the lowest score from the self-confidence sub-scale (37.89 ± 7.13).

Statistical no significant difference was determined among nurses' critical thinking disposition scores according to their working experiences, their working units ($p > 0.05$). However there was a statistical meaningful difference between the total score critical thinking dispositions of the nurses participate to service training activities ($p = 0.019$).

Conclusion: It was assigned critical thinking disposition scores of the nurses working at pediatric units are medium . However, participating in service training activities was determined to affect the critical thinking in a positive way.

1016

TETRAHYDROBIOPTERIN RESPONSIVENESS AFTER LOADING TEST OF 21 CZECH HPA/PKU PATIENTS AND CORRELATIONS TO THEIR GENOTYPE

D. Prochazkova¹, S. Pouchla², H. Vinohradská³, P. Konecna¹, Z. Dolezel¹

¹Department of Pediatrics, University Hospital Brno and Medical Faculty of Masaryk University, ²Center of Molecular Biology and Gene Therapy, ³Department of Biochemistry, University Hospital Brno, Brno, Czech Republic

Background and aims: Phenylketonuria (PKU;OMIM261600) is an inherited metabolic disease due to a deficiency of hepatic phenylalanin hydroxylase (PAH;12q24.1). Phenylalanine accumulation can lead to cognitive impairment. Tetrahydrobiopterin (BH4) responsiveness in patients with specific mutations in the PAH gene is a subtype of hyperphenylalaninaemia (HPA) characterised by a positive BH4 loading test.

Methods: We tested 21 patients with HPA/PKU, 4-39 years of age, selected (based on genotype) as a potentially BH4 responsive and loaded with BH4 (20mg/kg).

Results: Overall 9/21 showed positive response of more than 30% decrease in blood Phe levels 8 h after BH4 challenge, and 7/21 showed this decrease after 24 h. The majority of the responsive patients belong to mild HPA (11/16). Genotype analysis confirms the involvement in the response of specific mutations. In p. E390G, p.A300S, p. A403V, p.Y414C, p.I306V, p.G272X, p.I65T, p.R261Q and p.Y387H mutations were 100% associated with BH4 responsiveness. The p.R158Q mutation was inconstantly responsive. The EX5del-4kb mutation was responsive regardless of the second allele (p.A403V and p.R408W). In patient with mild PKU and novel mutation p.K396R (genotyp p.R408W/p. K396R) no responsiveness was noticed.

Conclusions: According to recent estimates, a group of individuals may benefit from BH4 substitution, eliminating the need of life-long dietary restrictios. The best responders were patients with mild HPA.