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

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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.

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

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Background and aims: Phenylketonuria (PKU; OMIM261600) is an inherited metabolic disease due to a deficiency of hepatic phenylalanine 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 inconsistently 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 (genotype 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 restrictions. The best responders were patients with mild HPA.