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Title: Sleep study characteristics in patients with mucopolysaccharidosis

Dr. Ayse Tana 3094 Aslan aysetana@yahoo.com MD [1], Dr. Cigdem 3095 Kasapkara ckasapkara@yahoo.com MD [2], Dr. Leyla 3096 Tumer ltumer@yahoo.com MD [2], Dr. Alev 3097 Hasanoglu Ahasanoglu@gmail.com MD [2] and Dr. Fatih 3098 Ezgu fezgu@yahoo.com MD [2]. [1] Pediatric Pulmonology, Gazi University, Ankara, Turkey and [2] Pediatric Metabolism, Gazi University, Ankara, Turkey .

Body: Background: Mucopolysaccharidoses (MPS) are inherited lysosomal storage disorders caused by the deficiency of hydrolases involved in the degradative pathway of glycosaminoglycans. In MPS, upper airway obstruction has multiple causative factors, may severely affect morbidity and mortality. Methods: We evaluated upper airway obstructive disease and related clinical findings through home sleep study in 19 patients (11 with MPS VI, 4 with MPS I, 4 with MPS II) with MPS followed at Gazi University Pediatric Pulmonology Unit. Patients underwent home based sleep measurements and sleep-respiratory problems were asked in a detailed clinical history. Measurements of apnea, apnea-hypopnea index, hypopnea index, oxygen desaturation index, minimal oxygen saturation were obtained through home sleep study Results: For 19 children, the disorder was normal in 1, mild (AHI 1.5-5/h) in 5, moderate (AHI 5-10) in 2, and severe (AHI>10) in 11. The prevalence of OSA was 94.7% (18/19) in patients with MPS. Snoring, witnessed apnea, pectus carinatum, and macroglossia were the main clinical findings. Echocardiograms showed evidence of pulmonary hypertension in 13 patients. Conclusion: Home sleep study is very quick, easily available screening test to determine the abnormalities of breathing during sleep and urge the clinicians to take necessary action for patients with severe manifestations.