Separation and Identification of Glycosaminoglycans in Urine of Patients with Mucopolysaccharidosis by Electrophoresis

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The Mucopolysaccharidoses (MPS) are a group of Lysosomal Storage Disorders caused by the deficiency of the enzymes that degrade glycosaminoglycans (GAGs) and consequently they are accumulated in the urine of patients. Currently, screening of this disease is made by measuring GAGs in the urine followed by chromatography to identify the type of GAG accumulation. The objective of this study was to standardize GAGs electrophoresis, separating and identifying these compounds increased in the urine of patients. For this, GAGs are extracted and purified from the urine sample of MPS I patients (5) and normal individuals (10) according Differrante (1967). The identification of GAGs is made by electrophoresis, based on the migration of different types of urinary GAGs: chondroitin, queratan, heparan and dermatan sulfate, in a discontinuous electric field on agarose gel, using also the properties of differential solubility in concentrations of ethanol. The staining is done with toluidine blue. Chondroitin and dermatan sulfate standards were applied. The results showed a band in the region of the pattern of chondroitin in urine of normal individuals and in patients samples with MPS I, a band of dermatan near the origin of electrophoresis application and a band between dermatan and chondroitin which is likely to heparan sulfate. This technique is very sensitive and can detect concentrations of up to three micrograms of GAGs. Moreover, the sharpness of the bands, the good reproducibility and reduced processing costs when compared to thin layer chromatography, makes of this technique an important tool for the diagnosis of MPS.

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