# Analysis of urine organic acids using Linear Retention Indices for the determination of inherited metabolic disorders

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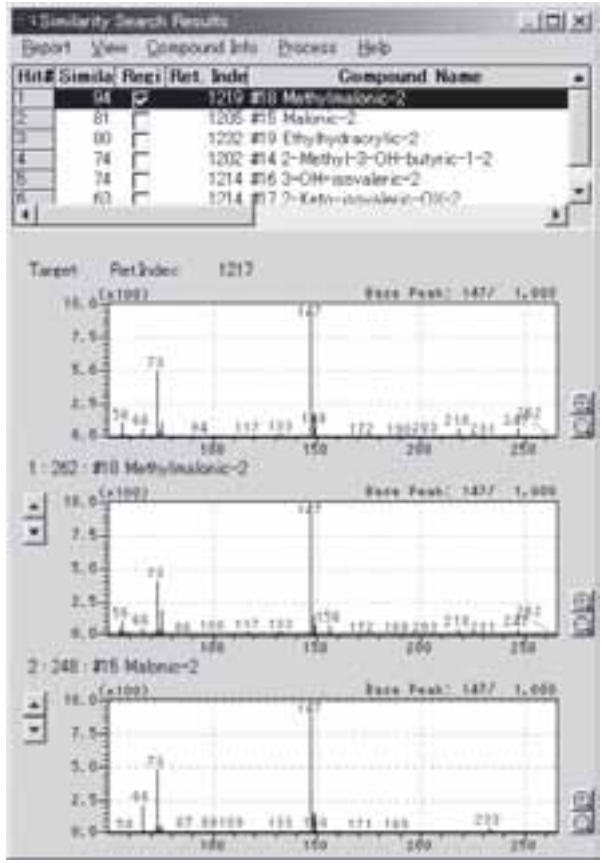
**AIMS:** Inherited metabolic disorders can be detected by the analysis of the accumulation of unmetabolized compounds in the human body. For this purpose the blood or urine of a patient is analysed for diagnostic markers with GCMS. In this application the analysis of organic acids in urine for diagnostic purposes for metabolic disorders is described using a new software with automatic calculation of Linear Retention Indices and an Excel table with threshold values developed together with Professor Yamaguchi of the Department of Pediatrics, Shimane University, Japan.

**METHODS:** Sample preparation: For sample preparation centrifuge the urine sample to remove the impurities then determine the creatinine content. Add urease solution to it, mix and let it react. Internal Standards are added to the urine sample. The pH is adjusted and an oximation reaction with hydroxyamine hydrochloride is performed. After acidification and extraction with ethyl acetate the sample is dried. Subsequently a mixture of BSTFA and TMCS for derivatization of the organic acids is added. GCMS analysis: The sample is then injected into the GCMS system. Identification of the urine organic acids is performed by using a database which contains the mass spectra and the Linear Retention Indices (LRI) of the organic acids. First a hydrocarbon standard is analyzed. Then the GCMS software (Shimadzu GCMSsolution) automatically calculates the LRI of the unknown compounds in the sample. In the subsequently library search not only the mass spectra of the peaks but also the LRI values of the peaks are compared with the database. This gives an unambiguous identification of the organic acids. Quantitative analysis is performed using the specific mass traces of the compounds.

**RESULTS:** For determination of the inherited metabolic disorders, qualitative and quantitative GCMS analysis of urine organic acids was performed and compared with threshold levels given in an Excel© table to link it with ca. 40 different corresponding disorders. The Excel table was developed together with Professor Yamaguchi of the Department of Pediatrics, Shimane University, Japan.Using this method metabolic disorders could be identified.




**KEYWORDS:** *Urine organic Acids; Linear retention Index; Metabolic disease*

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