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## 3-(5-methyl-1H-pyrazol-3-yl)propionic acid-d6 (MPP)

http://cn.lumiprobe.com/p/sa-succinylacetone-pyrazol-d6

Succinylacetone-pyrazole-d6 is a deuterated derivative of succinylacetone-pyrazole (SA-pyrazole-d6) and is used as an analytical standard in the measurement of succinylacetone (SA). Succinylacetone is a reactive diketone and reacts with the amino groups of amino acid residues of peptides and proteins present in the blood.

As a result, in order to extract the immobilized SA, it must be converted into a more highly stable product, which is subsequently subjected to extraction, namely a pyrazole derivative.

SA content in tandem mass spectrometry (MSMS) analysis is defined as the content of the derivative compound, 3-(5-methyl-1H-pyrazol-3-yl)propanoic acid (MPP). SA is quantified by adding a stable isotope-labeled propanoic acid analogue as an internal standard.

Succinylacetone is a product of tyrosine catabolism and an inhibitor of heme biosynthesis. Succinylacetone (SA) testing in dried neonatal blood spots, followed by quantitation of SA in blood or urine in high-risk neonates, has excellent sensitivity and specificity for the diagnosis of tyrosinemia type 1 [1]. Succinylacetone also serves as a tool for studying the defects of heme synthesis on cellular processes.

[1] Kehar M., Sen Sarma M., Seetharaman J., Jimenez Rivera C., Chakraborty P. Decoding hepatorenal tyrosinemia type 1: Unraveling the impact of early detection, NTBC, and the role of liver transplantation. Can Liver J. 2024. 7(1). P.54-63.