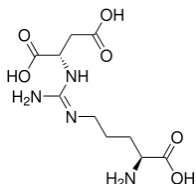


Separation of Argininosuccinic Acid on Biochrom Amino Acid Analysers

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Amino acid: Argininosuccinic Acid (ASA)



Instrumentation: Biochrom 20/20+/30/30+ Lithium Buffer System equipped with a High performance or high resolution Lithium column.

Method: optimised Lithium Accelerated, High Performance or High Resolution programme

Background:

Argininosuccinic aciduria is an inherited disorder that causes the accumulation of Argininosuccinic Acid (also known as "ASA") in the blood and urine. Some patients may also have an elevation of ammonia, which can affect the nervous system. Argininosuccinic aciduria may become evident in the first few days of life because of high blood ammonia, or later in life presenting with "sparse" or "brittle" hair, developmental delay, and

tremors. Argininosuccinic aciduria occurs in approximately 1 in 70,000 live births.

ASA tends to co-elute with Leucine on a standard chromatogram. This optimisation note describes how to modify the Biochrom standard programme in order to obtain full separation of ASA.

Methodology:

To improve the separation between ASA and Leucine, the time of the first step of buffer 3 (Lithium Citrate CII pH3.15) has to be increased until a satisfactory resolution is achieved between the two compounds. Proceeding by 1 minute increments is preferable. Overshooting the time of buffer 3 will cause ASA and Norleucine to co elute so a compromise has to be found.

When analysing ASA, it is recommended to use Aminoethyl Cysteine (AEC) instead of Norleucine as internal standard because the former elutes between Hydroxylysine and Ornithine. This gives more room for ASA to elute between Leucine and Tyrosine.

Physiological standard including Argininosuccinic Acid

