Rapid Analysis of Sulphocysteine

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Sulphite oxidase deficiency is a rare inborn error of metabolism, characterized by elevated urinary S-sulphocysteine. Using the Biochrom 30 Amino Acid Analyser, a short programme for the analyses of S-sulphocysteine has been developed.

Sulphite oxidase deficiency is a rare inborn error of the metabolism of sulphated amino acids, where sulphite oxidase activity is absent or diminished.

Individuals affected most commonly present in the neonatal period with severe seizures, characteristic dysmorphic features, and profound mental retardation.

Methionine and cysteine normally are metabolized to sulphite and then are oxidised to sulphate by the enzyme sulphite oxidase. When sulphate oxidase is deficient, alternate metabolic pathways for sulphite are augmented, including the formation of metabolites s-sulphocysteine and thiosulphate.

Determination of s-sulphocysteine in urine provides the most reliable indication of sulphite oxidase deficiency.

Analysis of sulphocysteine can be performed on a Biochrom 30 Amino Acid Analyser. The accurate quantification of this compound using a standard lithium high performance program can be difficult as it might not be well resolved from phosphoserine. A short program (53 minutes injection to injection) has been developed for the rapid quantification of sulphocysteine (S-cys) using D-Glucosaminic acid as the internal standard. The program is using lithium loading buffer pH 2.2 as buffer 1 instead of buffer A pH 2.8 and the temperature of injection is increased to improve the separation.

Reference

www.emedicine.com

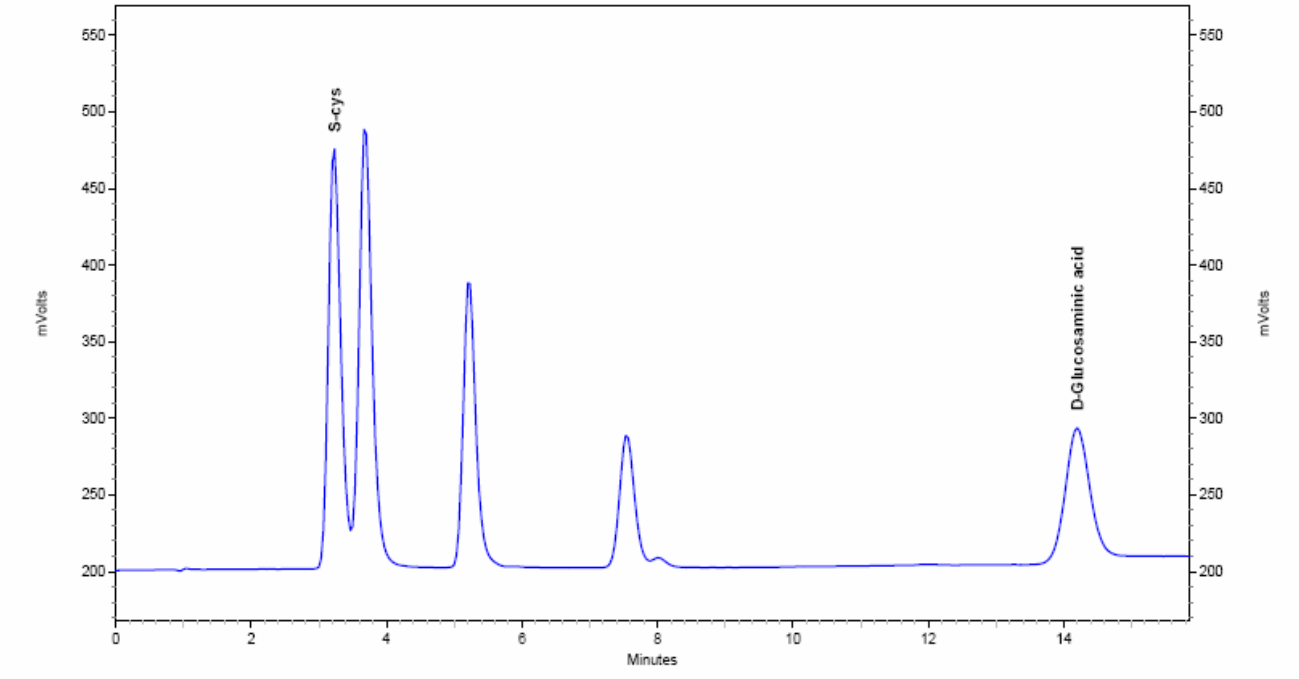

Figure 1. Short Program for the Analysis of Sulfocysteine using a Bio 30

BioSys

Sample: Physiological Fluid std

Amount Loaded: 10 nmol

Column Type: Peek

Column Number: U-1889

Resin Batch: 11956

Bed Length (mm): 200

Diameter (mm): 4.6

Instrument Serial Number: 93739

	Buffer	Nin
Flow Rate (ml/h):	30	20

	Buffer	**Molarity**	**pH**	**Batch No.**
Buffer 1 -	Lithium Loading buffer		2.20	12279
Buffer 2 -	Lithium Citrate Buffer B	0.30	3.00	12231
Buffer 6 -	Lithium hydroxide Solution	0.30		11728
Reagent	Ninhydrin			12296
	Ultrosolve			12307

Title: **Sulphocysteine**

Nin Flow Rate: 20.0 ml/h

No.	**Time**	**Temp**	**Buffer**	**Pump**	**Nin**	**Rec**	**Commands**
1	01:00	70°C	2	30.0ml/h	ON	OFF	
2	00:00	70°C	2	30.0ml/h	ON	OFF	Reset
3	01:00	70°C	2	30.0ml/h	ON	OFF	Load
4	06:00	70°C	2	30.0ml/h	ON	ON	
5	04:00	70°C	6	30.0ml/h	ON	ON	
6	06:00	70°C	1	30.0ml/h	ON	OFF	
7	30:00	70°C	1	35.0ml/h	OFF	OFF	
8	05:00	70°C	1	30.0ml/h	ON	OFF	