Product Data Sheet

Product Name: Tiopronin (Thiola)
Cat. No.: GC16209

Chemical Properties

Cas. No.	1953-02-2		
Chemical Name	2-(2-sulfanylpropanoylamino)acetic acid		
SMILES	CC(C(=O)NCC(=O)O)S		
Formula	$C_5H_9NO_3S$	M.Wt	163.19
Solubility	≥ 6.8mg/mL in DMSO	Storage	Store at -20°C
General tips	For obtaining a higher solubility , please warm the tube at 37 ℃ and shake it in the ultrasonic bath for a while.Stock solution can be stored below -20℃ for several months.		
Shipping Condition	Evaluation sample solution : ship with blue ice All other available size: ship with RT , or blue ice upon request.		
Structure	Tiopronin (Thiola)		

Background

Tiopronin (Thiola) is an inhibitor of cystine precipitation and excretion and used for the treatment of cystinuria [1].

Cystinuria is an inherited autosomal recessive disease that is characterized by the formation of cystine stones in the ureter, kidneys and bladder.

In seven cystinuric patients, tiopronin decreased the urinary supersaturation of cystine. The mean cystine capacity without tiopronin was 130.6 + 280.8, while the value during tiopronin use was 43.1 + 131.2 [1]. In 13 patients with cystinuria, the excretion of free cystine increased by 0.75 mg for each mM increase in urinary sodium, which suggested tubular reabsorption of cystine is in a sodium-dependent way [2]. In thirty-one patients with homozygous cystinuria, tiopronin reduced the urinary free cystine excretion by 0.15 mg in a dose-dependant way. Low doses of tiopronin increased urinary excretion of total cystine as well as the mixed disulfide [3]. In 13 patients with cystinuria, tiopronin decreased urinary cystine excretion from 901.48 mg to 488.60 mg [4].

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References:
[1]. Dolin DJ, Asplin JR, Flagel L, et al. Effect of cystine-binding thiol drugs on urinary cystine capacity in patients with cystinuria. J Endourol, 2005, 19(3): 429-432.
[2]. Lindell A, Denneberg T, Edholm E, et al. The effect of sodium intake on cystinuria with and without tiopronin treatment. Nephron, 1995, 71(4): 407-415.
[3]. Lindell A, Denneberg T, Jeppsson JO. Urinary excretion of free cystine and the tiopronin-cysteine-mixed disulfide during long term tiopronin treatment of cystinuria. Nephron, 1995, 71(3): 328-342.
[4]. Koide T, Yoshioka T, Miyake O, et al. Long-term study of tiopronin in patients with cystinuria. Hinyokika Kiyo, 2003, 49(2): 115-120.

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