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Cysteamine dose regimen

Received: 2 February 2006 / Accepted: 20 February 2006 / Published online: 1 June 2006
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Sirs,

In their article on a strict cysteamine dose regimen for children with cystinosis, Levtschenko et al. reported that the mean morning polymorphonuclear (PMN) cystine content after a 9-h-long interval following the evening cysteamine dose was significantly higher than that following a 6-h interval between doses [1]. These results are of clinical importance and are supported by J. Schneider et al. on plasma cysteamine concentration and PMN cystine content analysis [2].

On the basis of their data the authors concluded that patients should adhere strictly to a 6-hourly intake of cystagon. Assuming that a patient would take cystagon at 5-h intervals during the day, with a 9-h interval between the evening dose and the one the following morning, it is obvious that these patients would have a slightly higher PMN cystine content for 3 of the 24 h compared to those children taking cystagon exactly every 6 h. However, even with this higher PMN cystine content for these 3 h, the levels would remain below the critical value of 1 mmol/mg protein cystine.

The benefit of a constantly lower cystine level – achieved at the cost of sleep disruption – has to be balanced against patient non-compliance. Several groups have reported that non-compliance by patients with cystinosis may occur in almost 50% of the patients, mainly because of the side effects of cystagon. Superimposing a strict interval of a 6-hourly drug intake will interfere with sleep and may increase non-compliance even more.

References

1. Levtschenko EN, van Dael CM, de Graaf-Hess AC, Wilmer MJG, van den Heuvel LP, Monnens LA, Blom HJ (2006) Strict cysteamine dose regimen is required to prevent nocturnal cystine accumulation in cystinosis. *Pediatr Nephrol* 21: 110–113
2. Schneider JA, Clark KF, Greene AA, Reisch JS, Markello TC, Gahl WA, Thoene JG, Noonan PK, Berry KA (1995) Recent advances in the treatment of cystinosis. *J Inher Metab Dis* 18:387–397

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