Fällstrom, S. P., Lindblad, B., Lindstedt, S. and Steen, G. Hereditary tyrosinemia–fumarylacetoacetase deficiency. *Pediatr. Res.* 13 (1979) 78
Fällstrom, S. P., Lindblad, B. and Steen, G. On the renal tubular damage in hereditary tyrosinemia and on the formation of succinylacetoacetate and succinylacetone. *Acta Paediatr. Scand.* 70 (1981) 315–320
Fisch, R.O., McCabe, E. R. B., Doeden, D., Koep, L. J., Kohlhoff, J. B., Silverman, A. and Starzl, T. E. Homotransplantation of the liver in a patient with hepatoma and hereditary tyrosinemia. *J. Pediatr.* 93 (1978) 592–596
Furukawa, N., Kinugasa, A., Seo, T., Ishii, T., Ota, T., Machida, Y., Inoue, F., Imashuku, S., Kununoki, T. and Takamatsu, T. Enzyme defect in a case of tyrosinemia type I, acute form. *Pediatr. Res.* 18 (1984) 463–466
Glaser, J. H. and Sly, W. S. β-Glucuronidase deficiency mucopolysaccharidosis: methods for enzymatic diagnosis. *J. Lab. Clin. Med.* 82 (1973) 969–977
Goldsmith, L. A. Tyrosinemia and related disorders. In Stanbury, J. B., Wyngaarden, J. B., Fredrickson, D. S., Goldstein, J. L. and Brown, M. S. (eds.) *The Metabolic Basis of Inherited Disease*, McGraw-Hill, New York, 1983, pp. 287–299
Grenier, A., Lescault, A., LaBerge, C., Gagne, R. and Mamer, O. Detection of succinylacetone and the use of its measurement in mass screening for hereditary tyrosinemia. *Clin. Chim. Acta* 123 (1979) 93–99
Lindblad, B., Lindstedt, S. and Steen, G. On the enzymatic defects in hereditary tyrosinemia. *Proc. Natl. Acad. Sci. USA* 74 (1977) 4641–4645
Sassa, S. and Kappas, A. Hereditary tyrosinemia and the heme biosynthesis pathway: profound inhibition of δ-aminolevulinic acid dehydratase activity by succinylacetone. *J. Clin. Invest.* 73 (1983) 625–634
Tschudy, D. P., Hess, R. A., Frykholm, B. C. and Blaese, R.M. Immunosuppressive activity of succinylacetone. *J. Lab. Clin. Med.* 99 (1982) 526–532
Tuchman, M., Whitley, C. B., Ramnaraine, M.L., Bowers, L. D., Fregien, K. D. and Krivit, W. Determination of urinary succinylacetone by capillary gas-chromatography. *J. Chromatogr. Sci.* 22 (1984) 211–215
Wadman, S. K., Duran, M., Ketting, D., Bruinvis, L., Van Sprang, F. J., Berger, R., Smit, G. P. A., Steinmann, B., Leonard, J. V., Divry, P., Farriaux, J. P. and Cartigny, B. Urinary excretion of deuterated metabolites in patients with tyrosinemia type I after oral loading with deuterated L-tyrosine. *Clin. Chim. Acta* 130 (1983) 231–238
Weinberg, A. G., Mize, C. E. and Worthen, H. G. The occurrence of hepatoma in the chronic form of hereditary tyrosinemia. *J. Pediatr.* 88 (1976) 434–438
Weissmann, B. Synthetic substrates for α-L-iduronidase. *Meth. Enzymol.* 50 (1978) 141–150

J. Inher. Metab. Dis. 8 (1985) 24

Short Report

AN ARTIFACT IN URINARY AMINO ACID ANALYSIS PRODUCED BY TRIS(HYDROXYMETHYL)AMINOMETHANE (THAM)

W. G. Wilson[1,2] *and E. J. Squillaro*[2]

During amino acid analysis of urine from a patient treated with the organic buffer tris(hydroxymethyl) aminomethane (THAM), we found a large artifactual peak produced by that compound. Amino acids were analyzed by ion exchange using an automated D-300 amino acid analyzer system (Dionex, Inc.) with fluorometric detection and quantitation of amino acids following postcolumn derivatization with *o*-phthalaldehyde (OPA). A urine sample obtained after an exchange transfusion with THAM-buffered blood revealed a massive peak coeluting with phenylalanine. THAM, added to a control urine sample, produced a peak (retention time 91.1 min) corresponding to the unknown large peak (91.9 min). The peak area of THAM was only 7.5% as large as that of a comparable phenylalanine standard; the urinary THAM concentration in the patient was 446 200 μmol g^{-1} creatinine. Subsequent urine and plasma samples were normal.

THAM is an amino alcohol which is rapidly excreted by the kidneys (Nahas, 1962), and is sometimes used to treat patients with metabolic acidosis unresponsive to sodium bicarbonate. Although THAM was detectable using OPA fluorescence, a THAM spot on paper did not stain with ninhydrin. We wish to alert clinicians and biochemists to the possibility of a THAM-produced artifact in amino acid analyses using OPA.

[1] Corresponding author
[2] Department of Pediatrics, University of Virginia Medical Center, Charlottesville, VA 22908, USA

Reference

Nahas, G. G. The pharmacology of tris(hydroxymethyl) aminomethane (THAM). *Pharmacol. Rev.* 14 (1962) 447–472