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# Composition of Urinary Coproporphyrin Isomers I–IV in Human Porphyrias[1])

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*Dedicated to Professor Claude Rimington in honour of his 90th birthday*

**Summary:** The urinary distribution and relative proportions of the four coproporphyrin isomers I–IV were investigated in 50 patients suffering from hepatic and erythropoietic types of hereditary porphyrias. A highly efficient sample preparation method was applied to isolate urinary coproporphyrins, the isomer ratios of which were quantitated by isocratic ion-pair high-performance liquid chromatography. Results showed a significant decrease ($p < 0.001$) of the proportion of coproporphyrin I in acute hepatic porphyria (acute intermittent porphyria, hereditary coproporphyria, variegate porphyria, porphobilinogen synthase deficiency porphyria) as compared with chronic hepatic porphyria (porphyria cutanea tarda, chronic hepatic porphyria type B and C) (13.2 ± 5.3%, $\bar{x}$ ± S.D., vs. 31.4 ± 11.5%). Conversely, the proportion of isomer III was significantly higher ($p < 0.001$) in acute hepatic porphyria than in chronic hepatic porphyria (80.9 ± 5.2% vs. 62.2 ± 10.9%). As expected, the highest level of coproporphyrin I (90.0 ± 1.9%) was found in congenital erythropoietic porphyria.

The atypical coproporphyrins II and IV were detected in all types of porphyria analysed and ranged from 0.2 to 9.0%; no significant differences were seen between acute and chronic hepatic porphyrias. The diagnostic importance of the isomer ratios of coproporphyrins I and III has been confirmed in our study, while the significance of the atypical coproporphyrin isomers II and IV is still unclear at present.

## Introduction

Porphyrias are caused by deficiencies of haem biosynthetic enzymes (fig. 1). Characteristic excretion patterns of porphyrins and porphyrin precursors – reflecting the respective enzymatic defect – are observed in these conditions.

Differential diagnosis of porphyrias requires analysis of haem precursors in urine, feces, blood and tissues of affected patients (1, 2). Complementary studies of the haem biosynthetic enzymes in blood cells help to identify the nature of the hereditary defect (1, 2). Additional diagnostic information can be obtained for some porphyrias when the ratios of the naturally occurring isomers of series I and III are determined (3). Alterations of the normal ratios of coproporphyrin isomers I and III are found in cases of both hereditary and toxic porphyrias, e. g. lead poisoning (3–5). For instance, the well-known overproduction of type I isomers in congenital erythropoietic porphyria (4, 6) or the preponderance of heptacarboxyporphyrin III in porphyria cutanea tarda (3) greatly facilitates the biochemical diagnosis of these porphyrias. On the other hand, secondary copropor-

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