Role of the Kidneys in Porphyrrias

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Sir,

We read with great interest the recent article by Day et al. [1] regarding porphyrias and the kidney. We are now intensively concerned in our laboratory with the study of plasmatic porphyrins and porphyrinogens using direct fluorometry of diluted plasma [2,3].

Our results have shown the same representation of individual porphyrins in the plasma and morning urine of patients with porphyria cutanea tarda (PCT; fig. 1,2). A high content of oxidized porphyrins – uroporphyrin and heptacarboxyporphyrin – in plasma of PCT patients has been observed and we have shown a good correlation between the plasma level of uroporphyrin and heptacarboxyporphyrin and their urinary excretion in 24 h. This finding would imply an important role of the liver in PCT. We likewise proved that the excretion of various types of porphyrins in PCT varied according to a daily rhythm [4]. Uroporphyrin and heptacarboxyporphyrin, excreted in oxidized form [5], have a very similar rhythm, with the highest excretion rate in the early morning hours. Copro-porphyrin and pentacarboxyporphyrin, excreted as colorless porphyrinogens [5], also have very similar daily rhythms, with the highest excretion rates in the afternoon hours. However, the demonstrated rhythm of coproporphyrin and pentacarboxyporphyrin in PCT does not seem to play any important role in the total relationship between porphyrins excreted within 24 h. Nevertheless, our findings of different contents and a different representation of porphyrins in the liver and kidneys in a deceased patient with PCT (a 65-year-old man) as well as in 5 nonporphyric patients who died suddenly (table I) speak for a different synthesis of porphyrins in the liver and kidneys in PCT.

In lead poisonings, we have found a different representation of porphyrins in plasma and urine and likewise a different representation of porphyrinogens in plasma and urine (fig. 1,2). This difference makes us believe that porphyrinogens in lead poisoning are of renal origin. Sanguinetti et al. [6] pointed out that the highest levels of coproporphyrin in lead poisonings were excreted in the afternoon hours, as we have also demonstrated in PCT.

In variegate porphyria (VP) we have sometimes found high levels of porphyrins in plasma and almost undetectable levels of porphyrins in urine, while in other cases, especially in the acute attacks, we have found high levels of oxidized porphyrins in plasma and high levels of porphyrinogens (coproporphyrin and pentacarboxyporphyrin) in urine. Besides our findings, however, another picture can be seen in VP [1], i.e. low levels of plasma por-
Table I. Total porphyrins in the livers and kidneys of PCT patient and controls

Uro = Uroporphyrin; Copro = coproporphyrin.

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633 692 (nm)

Fig. 1. Fluorescence emission spectrum (excitation 397 nm, emission 570–700 nm) of diluted plasma from a patient with PCT (a) and from a patient with lead poisoning (b), with the corresponding thin layer chromatogram of methylesters of plasma porphyrins [7] and standard (st). U = Uroporphyrin; C = coproporphyrin; P = proto-porphyrin; RFI = relative fluorescence intensity.

Fig. 2. Spectral absorption of diluted fresh urine from a patient with PCT (a) and from a patient with lead poisoning (b) before and after oxidation (ox), with the corresponding thin layer chromatogram of methylesters of urinary porphyrins after oxidation [5].

In our opinion, the kidneys play an important role primarily in the synthesis of porphyrinogens, i.e. coproporphyrin and pentacarboxyporphyrin.

For a more detailed elucidation of the problem of porphyrin synthesis in the kidneys we consider important to examine the level of total porphyrins and the distribution of individual types of porphyrins in the liver and kidneys of patients with PCT before and after chloroquine therapy. Likewise, it will be useful to compare the levels and types of porphyrins in liver and kidney tissues of patients with chronic renal failure kept on long-term dialysis. Both these aspects are now under study on a greater number of patients.

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- kidneys and the liver in the synthesis of porphyrins in VP.