

SHORT COMMUNICATION

TREATMENT OF PHOTOSENSITIVITY IN CONGENITAL ERYTHROPOIETIC PORPHYRIA (CEP) WITH BETA-CAROTENE

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Abstract. Beta-carotene in combination with canthaxanthine has proved effective in a patient with CEP by preventing skin eruptions and increasing light tolerance as measured by graded light exposure with the Xenon lamp at 410 nm. It is important to administer a beta-carotene preparation which gives satisfactory serum levels (about 1000 $\mu\text{g}/100\text{ ml}$).

Recently we described a patient, a boy born July 1969, with the typical clinical picture of CEP (3, 4). The diagnosis was based on the clinical picture, an enormous increase of uroporphyrin and other porphyrins in the urine, the presence in the bone marrow of erythroblasts with fluorescence in the nuclei, and increased porphyrins also in red cells, plasma and feces. The porphyrin pattern of the patient, however, is in certain respects different from that reported in other cases of CEP, the main difference being that the isomer III porphyrins predominate, and not, as expected, the I isomers. The urinary excretion of large amounts of 7-carboxylic porphyrin is also exceptional. The patient is gravely affected with hemolytic anemia when not meticulously protected against daylight, and with photosensitivity of a very severe degree.

Stimulated by several recent reports on the effect of beta-carotene in increasing light tolerance and preventing skin eruptions in hereditary protoporphyria (1, 2, 5), we decided to try this drug on our patient. The purpose of this report is to present briefly the results of this therapeutic trial, which has been encouraging. To our knowledge beta-carotene has not been used in CEP before.

CASE REPORT

Details of the clinical picture have been reported earlier by Hofstad et al. (4). The patient's photosensitivity has been a great problem since he was about one year of age. Ointments have been of no help. Splenectomy, which may improve the situation in some cases of CEP, was performed in August 1972, but with no effect on the photosensitivity and hemolytic anemia. A therapeutic trial with large doses of vitamin E (up to 600 mg/day) was also unsuccessful. In order to avoid skin eruptions he had to be very carefully shielded against light of the shorter wavelengths. In his home, the window panes were covered with a yellow celluloid film (Para-Sol), which is not translucent for light with wavelengths below 510 nm. When outdoors, even in cloudy weather, he had to protect all parts of his body. He used a cowboy hat with yellow celluloid film hanging down as a shelter for his face and neck, and gloves on his hands. This meticulous light shielding became increasingly difficult to maintain as he grew older.

On July 4, 1973, treatment with special beta-carotene capsules with canthaxanthine, kindly supplied by Hoffmann-La Roche, was started. These capsules have proved valuable in obtaining high enough blood levels of beta-carotene in cases of hereditary protoporphyria (1). To begin with, one capsule containing 25 mg beta-carotene was given daily. The serum level of beta-carotene rose to 900 $\mu\text{g}/100\text{ ml}$ (upper normal limit 200 μg). Moderate carotene icterus developed. Under this treatment he improved markedly and could tolerate ordinary daylight considerably better, though direct sunshine on the skin has as far as possible been avoided. His hemoglobin level rose from 8.7 to 9.4 g/100 ml, despite the fact that he was much more exposed to light, and skin eruptions were few and mild. Light tests were performed with filtered light from an Osram High Pressure Xenon Arc Lamp (XBO 150 W) delivering 870 erg/sec/cm² at 410 nm. Before treatment he reacted with marked erythema after 10 min exposure, and with erythema and oedema after 20 min. Under treatment there was no reaction after 30 min exposure, only mild erythema after 40 min.

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