INSULIN SECRETION IN PATIENTS WITH MYOTONIC DYSTROPHY AND THEIR RELATIVES *

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Myotonia dystrophica is a hereditary disorder characterized by myotonia and muscular atrophy. Several endocrine and metabolic abnormalities in this disease have aroused interest including testicular atrophy, baldness, cataract, low basal metabolic rate and low 17-ketosteroids in the urine. Investigations of the endocrine function of the pituitary, thyroid and adrenal glands have failed to reveal any consistent abnormalities.

Huff et al. 7 observed elevated fasting plasma insulin concentrations in these patients and found that the insulin response to oral glucose was exaggerated. There is considerable disagreement, however, as to the consistency of this finding as well as glucose intolerance in this disorder. The present study was undertaken to further define the incidence of glucose intolerance and of hyperinsulinemia in patients with myotonic dystrophy and to determine whether such abnormalities occur in clinically unaffected close relatives of the patients.

MATERIALS AND METHODS

Eleven patients with clinically documented myotonic dystrophy and 9 of their unaffected close relatives were studied. Relatives were clinically free of disease, as assessed by history and physical examination. All were well nourished and had adequate carbohydrate intake prior to the study. Studies were performed in the morning on different days after overnight fasting. Healthy subjects, 20-45 years of age with

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normal body weight and without a family history of diabetes, were similarly studied as control subjects.

The dose schedules of various studies were: 1) 100 g of starch hydrolysate (Glucola®, Ames Company, U.S.A.), given orally; 2) 1 g i.v. tolbutamide given in 1 min; and 3) 5% arginine hydrochloride, 0.5 g/kg body weight, infused over 30 min.

Serum glucose was measured by the autoanalyzer procedure using the potassium ferricyanide reducing method. The University Group Diabetes Program criteria were used to assess glucose intolerance. Plasma immunoreactive insulin (IRI) was determined by coated charcoal immunoassay. The insulin responses were considered exaggerated when the maximum level attained exceeded the mean peak response of the control group by more than 2 SD.

RESULTS

Fig. 1 shows the glucose tolerance tests of 11 myotonic dystrophy patients, 9 unaffected relatives of the patients and 10 control subjects. Five myotonic dystrophy patients had glucose intolerance. IRI response of the 3 groups at various time intervals during the glucose tolerance test is shown in fig. 2. The mean fasting IRI levels were significantly elevated in the patient group (11.03 μU/ml for patients, 6.0 μU/ml for control groups, p<0.05). Seven patients, 4 of whom were diabetic, had hyperinsulinemic response to oral glucose and their peak insulin values ranged from 180-450 μU/ml (fig. 3). The mean peak insulin concentration in myotonic dystrophy patients was significantly higher than in control groups.