PROGRESSIVE SYSTEMIC SCLEROSIS (DIFFUSE SCLERODERMA)

TREATMENT WITH POTABA—A PRELIMINARY REPORT

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Progressive systemic sclerosis or diffuse scleroderma is a disease of the connective tissue of unknown aetiology. No system of the body is immune in the disease process and the course is variable and unpredictable. In its fully developed form the disease produces severe disability and is not uncommonly associated with a high mortality rate.

The purpose of this preliminary communication is to describe a case of diffuse scleroderma which has responded to Potaba (potassium p-aminobenzoate Glenwood). The beneficial effect of this drug was first described by Zarafonetis in 1950.[1]

Case Report

Mrs. K.A., a 46-year-old White female, had in 1953 developed Raynaud's phenomenon involving the fingers and toes of all the limbs, and which was always precipitated by cold weather and associated with tightness of the skin.

The diagnosis of scleroderma was made and she was treated with ACTH which, however, produced hypoglycaemic episodes. Cortisone thereafter precipitated hypercortism. In 1956, at the Peripheral Vascular Clinic, Groote Schuur Hospital, it was found that she had Raynaud's phenomenon of the hands and feet, associated with gangrenous patches of the fingertips and toes, and cutaneous scleroderma involving the upper and lower limbs, trunk, face, and neck. There was marked limitation of the large and small joint movements and gross reduction in the opening of the mouth.

The blood count showed a haemoglobin of 12·5 G and the sedimentation rate was 10 mm. in the 1st hour (Westergren). Barium studies and limb X-ray examination was normal. She was again treated with cortisone with only intermittent effect.

She was admitted to the surgical wards in 1957 for quadruple sympathectomies. Postoperatively all the symptoms persisted and later became aggravated.

In 1959, with the cutaneous and joint symptoms still evident, she developed gastro-intestinal symptomatology, viz. epigastric postprandial discomfort, heartburn, and constipation. Various courses of therapy were instituted with no lasting effect.

In 1964 her joint stiffness was incapacitating, the cutaneous scleroderma involved the whole body, and Raynaud's phenomenon was gross and associated with tapering and ulceration of the digits. Dysphagia was very marked in addition to heartburn, epigastric pain and constipation.

A barium meal showed marked incoordination of oesophageal peristalsis, motility abnormality of the stomach, small

intestine and large bowel. There was in addition gross scarring of the duodenum.

Augmented histamine test: Basal secretion 4·7 mEq. and the maximum acid output was 40·5 mEq.

Potaba was prescribed as a last resort in November 1964 since the patient had already threatened suicide and because of the progress and deterioration of the patient's physical condition. The response was phenomenal. In March 1965 the skin texture had improved, the mouth was mobile, joint movements had remarkably improved and the gastro-intestinal symptoms had practically abated except for the duodenal ulcer symptoms which, however, were responding slowly to an ulcer regime. She now felt normal again.

COMMENT

The purpose of this case report is to indicate to general practitioners that we are conducting a trial with this drug in the treatment of diffuse scleroderma, and would appreciate it if all cases of diffuse scleroderma could be referred to the Peripheral Vascular Clinic, Groote Schuur Hospital, by appointment, for investigation and treatment.

We wish to thank Dr. J. G. Burger for permission to publish this case report, and Protea Pharmaceuticals for the generous supply of Potaba.

REFERENCE

1. Zarafonetis, C. J. D. *et al.* (1950): Arch. Intern. Med., **85**, 27.