

in July 1971, another (48) has not been seen since October 1967, and three (8, 13, and 24) had emigrated in good health in 1968, 1969, and 1971, respectively. Eleven patients had died. The table shows the actual number of deaths compared with the expected number in 5-year age groups. Cerebrovascular disease probably caused death in four cases, and cardiovascular disease in two. Two others died suddenly and unexpectedly while recovering from a severe painful crisis and the lack of causal pathology at necropsy was compatible with a cardiac arrhythmia. Forty two were traced during 1973, of whom 32 had regularly attended the clinic during the past five years, six had been specially contacted and examined for the present survey, and four (2, 22, 40, and 54) had not been seen but had corresponded or were reliably reported alive. Thirty three patients were in reasonable health, 22 of them in full time employment. In nine cases, symptoms of the disease interfered with normal activity, owing to leg ulceration in six, low haemoglobin levels in two, hepatic enlargement and abdominal swelling in one.

When analysed in 5-year age groups the haematological characteristics of the elderly group did not differ significantly from those of younger ages.

## Discussion

Prolonged survival in S.S. disease does not mean a benign process. In only two patients was it diagnosed incidentally and these admitted to mild symptoms on direct questioning. Most patients had experienced typical and occasionally severe symptoms of S.S. disease when younger. After the age of 30, painful crises tended to become less frequent and less severe, leg ulcers healed, and aplastic crises were less common. Despite this symptomatic amelioration, however, vessel obstruction and ischaemia progressed. Occlusion of the peripheral retinal vessels resulting in retinitis proliferans occurred in 7/20 (35%) of patients aged over 48 compared with 1/30 (3%) of those under this age. Abnormalities of renal function also increase with age.

There is now good evidence that S.S. disease is compatible with survival into the fourth, fifth, and sixth decades though increased mortality, characteristic at younger ages, persists. There is a lower proportion of elderly patients attending the clinic than would be expected had the condition been benign. This might result from either a high mortality or from the lack of diagnosis in the elderly. Our evidence confirms the high mortality, which was eleven times that expected in the general Jamaican population. Late presentation is also a contributing factor, however, since 57 patients over the age of 30 including 10 over 50 have joined the clinic during the 5-year follow-up.

It is not understood why some patients survive to comparatively old ages and others die young. The role of other genetic and environmental factors<sup>5</sup> remains speculative. So also does the natural history of the disease. A prospective study from birth is the only way of obtaining an accurate assessment of the natural history of the disease and would also allow the influence of environmental and genetic factors to be assessed.

I wish to thank Dr. Michael Ashcroft of the M.R.C. Epidemiology Unit for the calculations of expected deaths in the Jamaican population.

<sup>1</sup> Serjeant, G. R., *et al.*, *British Medical Journal*, 1968, 3, 86.

<sup>2</sup> Ali, S. A., *British Journal of Haematology*, 1970, 19, 613.

<sup>3</sup> Gelpi, A. P., *et al.*, *Acta Haematologica*, 1970, 43, 89.

<sup>4</sup> Perrine, R. P., *et al.*, *Lancet*, 1972, 2, 1163.

<sup>5</sup> Steinberg, M. H., *et al.*, *Journal of the American Medical Association*, 1973, 224, 317.

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## Method of Using Pressurized Aerosols

The efficacy of inhaled bronchodilators depends on demonstrating the correct method to the patient. The timing of the inhalations is important when the inhaler is taken according to the manufacturer's instructions. The inhaler appears to be far more effective if the aerosol is released at some distance from the open mouth rather than directly into it—for two reasons: firstly, a larger fraction of the dose from the aerosol would be impacted on the back of the palate when the aerosol was discharged at a close range (that is, closed mouth); and, secondly, better entraining of the aerosol with the inspired air

parallel to but wider than the aerosol jet. A single blind method was used to test this hypothesis in terms of improving the patient's peak expiratory flow rates (PEFR) and preliminary results confirm this.

## Materials and Methods

Thirty one patients with airway obstruction, which was reversible by at least 10%, inhaled isoprenaline. Most patients were familiar with using inhaled bronchodilators. The peak flow rate was measured by the standard Wright peak flow meter. Isoprenaline was delivered by two puffs of the standard Medihaler Iso, which contained 0.1 mg of isoprenaline. Patients were instructed to use the inhaler at a distance of 40 mm from the open mouth with a blank (open mouth). Their technique with the orthodox (closed mouth) was also confirmed. They were randomly allocated to two groups and their response to isoprenaline inhaler was followed according to the protocol below:

	GROUP A	GROUP B
	Control PFR	Control PFR
Time 0 min.	Inhale with open mouth	Inhale with closed mouth
5 "	PEFR	PEFR
10 "	Inhale with closed mouth	Inhale with open mouth
15 "	PEFR	PEFR

Three patients attended one week later, when they were put in the opposite group. A doctor allocated the patients to treatment groups and instructed them. Peak flow readings were recorded by a technician, who was unaware of the patient's group. The initial peak flow was best of three readings and subsequent peak flows the better of two readings. Readings were converted to percentages of the control PEFR (see table).

### Peak Flow Rates in Patients Inhaling with Open (A) and Closed (B) Mouth

Group	Control Peak Flow		Initial Improvement		Further Improvement	
	Range	Mean	Range	Mean	Range	Mean
A	60-400	195	7-78	30.2	-9 + 23	3.9
B	70-420	212	-2 + 40	14.5	-1 + 29	10.1

Groups A and B had the same mean peak flow measurement before treatment. The mean percentage initial improvement in PEFR was significantly higher in Group A with mouth open than in Group B with mouth closed ( $t=2.83$ ;  $P<0.05$ ). The mean percentage further improvement in Group B with mouth open was also significantly greater than in Group A with mouth closed ( $t=2.21$ ;  $P<0.05$ ). The overall improvement in PEFR after both treatments (open + closed mouth) did not differ significantly between A and B ( $P>0.05$ ).

## Discussion

With the Medihaler, as used in these experiments, impaction judged by wetting sheets of paper no longer occurs at approximately 11 cm. The delivery of the standard salbutamol (Ventolin) and steroid inhalers (Bextosol and Becotide) appear to be more powerful. The impaction distance is up to 22 cm with Ventolin and rather less with the steroid preparations. In all cases this distance is far longer than the distance from the mouthpiece to the soft palate when using the closed mouth method. That most of the aerosol is not entrained is confirmed by the fact that most of the dose is swallowed.<sup>1-3</sup> The distribution of thrush (the most common complication with steroid inhalers) on the soft palate suggests that impaction is an important factor. Moreover, limited personal experience suggests that if the open mouth method is used thrush is less persistent with high doses of steroid inhaler, and the lesions are smaller and more widely distributed in the mouth, presumably because of less impaction on the soft palate.

I wish to thank Mr. R. A. McNay, statistician, Northern Regional Health Authority, for performing the statistical analysis.

<sup>1</sup> Blackwell, E. W., *et al.*, *British Journal of Pharmacology*, 1970, 39, 194.

<sup>2</sup> Evans, M. E., *et al.*, *British Journal of Pharmacology*, 1971, 43, 466.

<sup>3</sup> Walker, S. R., *et al.*, *Clinical Pharmacology and Therapeutics*, 1972, 13, 861.

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