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Upper airway obstruction complicating the Shy-Drager syndrome

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Paralysis of the laryngeal abductor muscles leading to stridor is a rare feature of the syndrome of progressive autonomic failure with multiple system atrophy (Shy-Drager syndrome).[1,2] We report a patient with the Shy-Drager syndrome and abductor paralysis in whom the airway narrowing appeared to be partly dynamic and in whom treatment with levodopa produced a demonstrable improvement in airflow.

Case report

A 38 year old man presented in 1978 with tremor, impotence, postural syncope, and severe constipation. Examination confirmed a Parkinsonian tremor, generalised rigidity, and bradykinesia. Appreciable postural hypotension was also noted. A diagnosis of Shy-Drager syndrome was made and the patient was treated with levodopa and carbidopa (Sinemet), ephedrine, and sodium chloride. He was able to continue work as an electronics engineer and apart from gradual worsening of the Parkinsonian symptoms he remained well for the next three years.

In 1981 he complained of a weak voice and his wife noted nocturnal snoring. There were no other symptoms referable to the respiratory system. Indirect laryngoscopy showed bilateral abductor paralysis with an effective aperture of about 4 mm. Over the next three months he complained of increasing breathlessness, and he spontaneously mentioned improvement in the respiratory symptoms about one hour after treatment with levodopa.

In March 1982 he was referred for assessment of respiratory function. At that time he was able to walk more than 300 metres on the level. His voice was high pitched and he was unable to shout; he had considerable inspiratory stridor when talking and on even slight exertion. Inspiratory indrawing of the sternum was noted and this paradoxical movement was confirmed with magnetometers. Spirometry showed normal FEV_1, vital capacity, lung volumes, and carbon monoxide transfer. Specific airways conductance (sGaw) was reduced at 0·084 cm H_2O^{-1} s^{-1} (normal range 0·13 – 0·35 cm H_2O^{-1} s^{-1}). The maximum flow volume curve (fig 1) showed a pattern consistent with severe extrathoracic airway obstruction with particularly marked limitation of maximum inspiratory flow ($\dot{V}_I$max).

Over the subsequent six months the patient's condition deteriorated—his exercise tolerance worsened and the snoring began to disturb his neighbours. He was, however, still actively employed. He had no morning headaches or daytime somnolence. He was reassessed in August 1982, when physical examination showed no change. The electrocardiogram and chest radiograph remained normal. Arterial oxygen tension was normal at 11·6 kPa (87 mm Hg) but there was mild hypercapnia ($PaCO_2$ 6·3 kPa; 47 mm Hg). Flow volume curves showed worsening flow rates and there was a deterioration in the results of spirometric tests. Sleep monitoring over six and a half hours showed three periods of central apnoea but no obstructive apnoea; paradoxical motion of the ribcage was exaggerated and associated with audible inspiratory stridor.

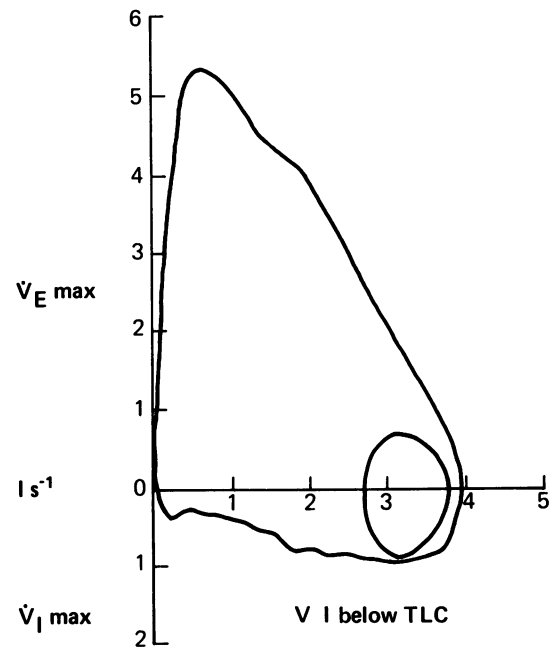

Fig 1 *Maximum and tidal flow volume curves. There is a major disparity between maximum expiratory and inspiratory flow rates. Close proximity of tidal and maximum inspiratory flow rates is also shown. V—volume; $\dot{V}_E$max, $\dot{V}_I$max—expiratory, inspiratory, maximum flow.*

EFFECTS OF TREATMENT

A double blind comparison of levodopa and benserazide (Madopar 250) with identical placebo showed improve-

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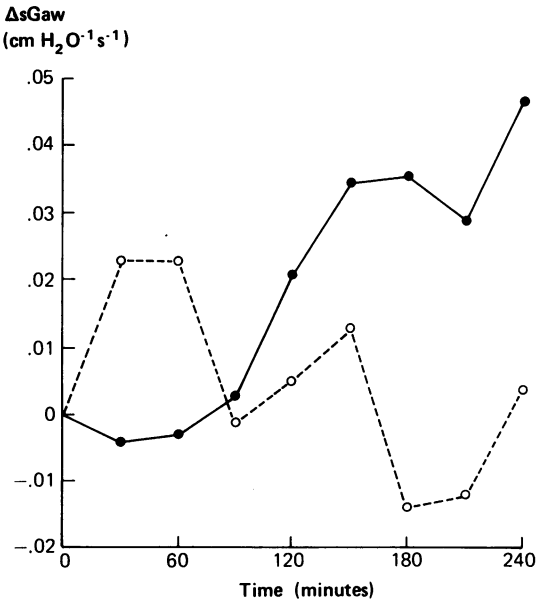

Fig 2 *Changes in sGaw (ΔsGaw) after placebo (open circles) and levodopa/benserazide (Madopar) (closed circles) in relation to predrug values. Improvement was seen two to four hours after the active preparation had been taken.*

ment in sGaw, which was measured at 30 minute intervals, after the active drug (fig 2).

Despite subjective and objective evidence of improvement with treatment, airway function remained severely compromised. A tracheostomy was therefore performed, after which his exercise tolerance improved dramatically. Six months later his main complaint was of worsening episodes of akinesia not responsive to levodopa.

Discussion

The finding of adducted vocal cords with paralysis of the abductor apparatus has recently been recognised as a complication of Shy-Drager syndrome. Although it appears to be rare, the series of Williams *et al*[1] suggests that it may not be as uncommon as was previously believed and should be looked for carefully in such patients. Previous reports[1 2] have commented on two features not seen in this case—oedema of the vocal cords and surrounding folds and the presence of obstructive sleep apnoea.[3] Israel and Marino[4] reported a patient with Shy-Drager syndrome who needed a tracheostomy for upper airway obstruction. In that case the flow volume curve was said to show an "inspiratory pattern typical of upper airway obstruction" but no further details were given. Our patient had severe upper airway obstruction demonstrated by means of flow volume curves. A disparity between expiratory and inspiratory flow is characteristic of dynamic or variable obstruction of the airway[5] but the disproportionate reduction of $\dot{V}_I$max in this patient was greater than is usually seen with bilateral vocal cord paralysis; the decreased sGaw indicates structural narrowing as well and is consistent with the appearances on laryngoscopy. A possible mechanism for the disproportionately reduced $\dot{V}_I$max in our patient is a combination of decreased tone in the muscles of the upper airway and an unusually negative airway pressure during inspiration. The reduced specific conductance measured during panting at low flows will therefore tend to underestimate the airway narrowing occurring during more forceful inspiratory efforts.

Upper airway obstruction in patients with extrapyramidal lesions is not confined to the Shy-Drager syndrome. Vas *et al*[6] reported two patients with Parkinsonism but no autonomic features who developed "laryngeal spasm" and one of these died from airway obstruction. The other patient improved after tracheostomy. To our knowledge no previous report of the effect of levodopa treatment on this manifestation of the Shy-Drager syndrome or Parkinson's disease has been reported. In the patient described here levodopa produced some improvement in upper airway function but this was insufficient to avoid tracheostomy, which has completely relieved the patient's respiratory symptoms.

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