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Neurological Improvement Following Respiratory Support in Syringomyelia: Case Report

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Summary

Respiratory failure in syringomyelia has rarely been reported. We report a patient with syringomyelia who showed some neurological improvement after intermittent respiratory support with negative and positive pressure ventilation.

Key words: *Syrinx, Syringomyelia, Respiratory insufficiency, Hypercapnea.*

Syringomyelia is a chronic disorder characterised by the presence of a cavity, surrounded by gliosis in the spinal cord or brainstem (Walton, 1985). Rarely, it has been reported to be associated with respiratory insufficiency (Nognes *et al.*, 1988; Bokinsky *et al.*, 1973, Haponik *et al.*, 1983, Weiner, 1980).

We report a case of a patient with syringomyelia who demonstrated marked improvement in respiratory and voice function after negative pressure ventilation and the use of a positive pressure pneumobelt.

Case report

A 43-year-old man was admitted to our rehabilitation hospital for adaptive equipment and pulmonary evaluation. The patient had a 20-year history of syringomyelia which had been previously diagnosed by myelography. The syrinx involved both the high cervical spinal cord and brainstem, and probably communicated with the ventricular system. For the last 10 years the patient had no useful upper extremity function, very limited lower extremity function, and suffered from dysphagia. He had multiple hospitalisations for aspiration pneumonias.

The patient was known to be hypercapnic for at least 6 months prior to his admission to our facility. He complained of morning headaches, periorbital swelling and he was sleeping poorly. His voice volume had progressively decreased over the previous 10 years. Six months prior to admission the vital capacity was 870 cc, and the arterial blood gas was pH 7·41 mmHg, PCO_2 46 mmHg, PO_2 64 mmHg.

Physical examination revealed a cooperative, pleasant white male with shallow respiration and weak voice, with a pulse of 65 per minute and regular, a blood pressure of 120/60, a respiratory rate of 25, and a temperature of 97·5° degrees Fahrenheit. His general physical examination was significant for paradoxical breathing and decreased breath sounds at the right base. His neurological examination was remarkable for a left 6th nerve palsy, nystagmus

in lateral gaze, and right tongue atrophy. Motor strength was assessed descriptively and by Medical Research Council criteria (Medical Research Council, 1982). Upper extremity strength proximally was 0/5. In the distal upper extremity there was trace movement. In lower extremity there was 3/5 proximal lower extremity strength with bilateral foot drop 2/5 and increased tone. No peroneus longus or brevis power was noted 0/5. Pin prick was decreased over the left side of the body. There were bilateral Babinski signs.

On admission the haemoglobin was 16·0 g/dl, haematocrit 47·4% and arterial blood gas was pH 7·37 mmHg, PCO_2 62 mmHg, PO_2 55 mmHg. Nasal cannula 1 litre O_2, arterial, blood gas was pH 7·36 mmHg, PCO_2 64 mmHg, PO_2 92 mmHg, vital capacity was 200 cc.

The patient was evaluated by oximetry. Oxygen saturation decreased intermittently to 60% during sleep. No periods of sleep apnea were observed.

He was treated with a negative pressure respirator cuirass at night with a setting of 32 cm H_2O and respiratory rate 18 per minute. A positive pressure pneumobelt was used for several hours during the day with settings of +30 cm H20 and rate of 15 per minute. The pneumobelt is an exsulflation belt that is fitted around the waist of a patient. It is attached to a ventilation source that inflates the bladder at adjustable rate and pressure setting, so as to augment diaphramatic breathing.

On this regimen the patient subjectively felt stronger with a louder and more sustained voice. The morning headaches and periorbital swelling decreased. His vital capacity increased to 800 cc and his day arterial blood gases with no respiratory device improved to pH 7·40 mmHg, PCO_2 57 mmHg, PO_2 71 mmHg on room air. Whereas previously right thumb adduction was only trace, right thumb improved to weak but nearly full adduction. The left peroneus longus and brevis improved to 2/5. These movements were not noted prior to treatment with negative pressure ventilation.

Discussion

There are two possible mechanisms to explain the patient's improved respiratory status with the use of regular night time negative pressure ventilation followed by several hours of the positive pressure pneumobelt during the day. One possibility is that by simply resting the respiratory muscles they are able to work more effectively when they are not being assisted mechanically. However, this would not explain the possible small, but significant improvements in motor function in the right thumb and left ankle.

A possible hypothesis to explain increased respiratory capacity and increased motor function is the following: reduction in PCO_2 may have lead to decreased pulmonary artery pressures, which translated into a generalised lowering of venous pressure (Austin *et al.*, 1957). Lower intracranial venous pressure in turn leads to decreased cerebral spinal fluid pressure (Harrison, 1934; Friedfeld *et al.*, 1934). Lower cerebral spinal fluid pressure within a syrinx may have caused alleviation of pressure on critical spinal and brainstem structures leading to improved neurological function (Gardner and Angel, 1958).

Our case suggests that in patients with syingomyelia and respiratory insufficiency aggressive treatment of hypercapnea may serve as a useful adjunct to standard neurological and neurosurgical management that directly addresses the underlying structural abnormality.

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