

ledge only one patient has been reported with an association between obstructive sleep apnoea proven by a sleep study and adult acquired micrognathia due to rheumatoid arthritis.<sup>3</sup> However, in that case there was no upper airway imaging. Our patient showed an extremely reduced upper airway area (21 mm<sup>2</sup> and 68 mm<sup>2</sup> at the oropharyngeal and hypopharyngeal levels, respectively) which may have been a major factor in the collapse of the upper airway.

The destruction of the cervical spine by rheumatoid arthritis, leading to potential compression of the cord or medulla, has been advocated as a causative factor for central sleep apnoea.<sup>3</sup> Although our patient had an atlanto-axial dislocation and a subluxation of C3-C4, she had predominantly obstructive apnoeas, a normal ventilatory response to carbon dioxide, and no brainstem compression on the CT scan. This mechanism is therefore unlikely to explain the sleep apnoea syndrome in this case.

Sleep fragmentation in rheumatoid arthritis with marked disruption of sleep continuity<sup>7</sup> can occur without sleep apnoeas as a result of pain, periodic leg movements, depression, and effects of drugs. Sleep fragmentation could favour respiratory instability and sleep apnoea including obstructive events.

In the present study, however, the abnormalities of the upper airway can reasonably be considered to be the main cause of the obstructive sleep apnoeas.

In clinical practice it is important to consider the diagnosis of sleep apnoea in patients with rheumatoid arthritis with temporomandibular joint destruction or cervical spine lesions who present with sleepiness, snoring, and/or cardiovascular disease. Prospective studies to assess the incidence and causative factors of sleep apnoea in large groups of patients with rheumatoid arthritis are needed.

- 1 Pépin JL, Lévy P, Veale D, Ferretti G. Evaluation of the upper airway in sleep apnea syndrome. *Sleep* 1992;15: S50-5.
- 2 Davies SF, Iber C. Obstructive sleep apnea associated with adult-acquired micrognathia from rheumatoid arthritis. *Am Rev Respir Dis* 1983;127:245-7.
- 3 Fisher MA, Casey LC, Ellman MH, Perlick SJ. Sleep apnea due to odontoid brainstem compression in a patient with rheumatoid arthritis (abstract). *Neurology* 1986;36(Suppl 1):163.
- 4 Rechtschaffen A, Kales A. *A manual of standardized terminology, technique and scoring system for sleep stages of human sleep*. Brain Information service, Brain Information Institute, University of California, Los Angeles, 1968.
- 5 Riley R, Guilleminault C, Herran J, Powel N. Cephalometric analyses and flow-volume loops in obstructive sleep apnea patients. *Sleep* 1983;6:303-11.
- 6 Pépin JL, Ferretti G, Veale D, Romand Ph, Coulomb M, Brambilla C, et al. Somnofluoroscopy compared with computerised tomography of the airway and cephalometry in obstructive sleep apnoea. *Thorax* 1992;47:150-6.
- 7 Mahowald MW, Mahowald ML, Bundlie SR, Ytterberg SR. Sleep fragmentation in rheumatoid arthritis. *Arthritis Rheum* 1989;32:974-83.
- 8 Lavie P, Nahir M, Lorber M, Scharf Y. Nonsteroidal anti-inflammatory drug therapy in rheumatoid arthritis patients. Lack of association between clinical improvement and effects on sleep. *Arthritis Rheum* 1991;34:655-9.
- 9 Young T, Palta M, Dempsey J, Skatrud J, Weber S, Badr S. The occurrence of sleep-disordered breathing among middle-aged adults. *N Engl J Med* 1993;328:1230-5.
- 10 Redlund-Johnell I. Upper airway obstruction in patients with rheumatoid arthritis and temporomandibular joint destruction. *Scand J Rheumatol* 1988;17:273-9.

*Thorax* 1995;50:694-695

## Paradoxical vocal cord adduction in an adolescent with cystic fibrosis

P Shiels, J P Hayes, M X FitzGerald

### Abstract

**Many patients with cystic fibrosis have symptoms of dyspnoea and wheeze which are responsive to treatment with bronchodilators. An adolescent woman with cystic fibrosis is described who presented with inspiratory stridor and in whom the classical features of paradoxical vocal cord adduction were found.**

(*Thorax* 1995;50:694-695)

**Keywords:** cystic fibrosis, vocal cord adduction, airways obstruction.

Paradoxical vocal cord adduction is a very rare functional disorder caused by inappropriate adduction of otherwise normal vocal cords. Typically, it affects women under 40 years of age who often have a background of employment in health care. The upper airways obstruction associated with this condition may be mistaken for asthma and symptoms may persist in spite of varying therapeutic interventions. We describe here an adolescent woman with cystic fibrosis with the classical features of paradoxical vocal cord adduction – an association, to our knowledge, not previously described.

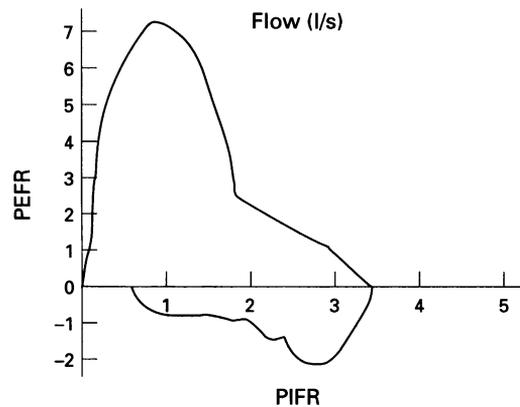
### Case report

A 17 year old adolescent woman was admitted in July 1993 for assessment of chronic, predominantly dry, daytime and nocturnal cough with minor hoarseness. Cystic fibrosis was diagnosed at birth and she attended the Adult Cystic Fibrosis Unit at St Vincent's Hospital, Dublin from 1991. Features of her illness included chronic bronchiectasis, multiple nasal polyps, pancreatic insufficiency, and recurrent meconium ileus equivalent. Her sputum was

Department  
of Respiratory  
Medicine and Cystic  
Fibrosis, St. Vincent's  
Hospital/University  
College Dublin,  
Dublin 4,  
Ireland  
P Shiels  
J P Hayes  
M X FitzGerald

Reprint requests to:  
Professor M X FitzGerald.

Received 9 March 1994  
Returned to authors  
2 June 1994  
Revised version received  
21 September 1994  
Accepted for publication  
26 September 1994



Flow-volume loop showing severe inspiratory obstruction.

chronically colonised with *Pseudomonas aeruginosa*. In 1992 she was admitted with newly acquired symptoms of wheeze and cough. At that time she was found to have a significant reduction in forced expiratory volume in one second (FEV<sub>1</sub>) of 1.77 l (50% pred) with a vital capacity (FVC) of 2.71 l (73% pred) and an 18% improvement in FEV<sub>1</sub> after inhalation of salbutamol, consistent with a diagnosis of asthma. She was commenced on inhaled corticosteroids, inhaled bronchodilators, and oral theophylline. This resulted in considerable improvement in her symptoms of wheeze but the cough persisted. In the summer of 1992 she first developed prominent hoarseness and noisy sounds in her throat. There was no evidence of oropharyngeal candidiasis or urticaria. The addition of oral corticosteroids did not result in any clinical improvement and she was admitted for assessment. Following admission she was noted to have considerable inspiratory stridor with minimal expiratory wheeze. Lung function measurements of expiratory volumes and flow rates were surprisingly normal with an FEV<sub>1</sub> of 3.05 l (95% pred), FVC 3.65 l (101% pred), and a carbon monoxide transfer factor (TLCO) of 23.18 (78% pred). However, examination of the flow-volume loop showed significant persistent inspiratory upper airways obstruction (figure). Considerable paradoxical adduction of the vocal cords was found on bronchoscopy carried out under local anaesthesia. She was prescribed codeine phosphate linctus with symptomatic relief. Initial assessment by the clinical psychologist attached to the unit confirmed the patient's desire to pursue a career in health care but did not reveal any psychological abnormality. Long term psychological assessment was declined by the patient's family.

### Discussion

Paradoxical vocal cord adduction is an unusually rare condition due to inappropriate adduction of the vocal cords.<sup>1</sup> It usually affects

young adult women working in health care-related jobs. While functional airways obstruction is most often the result of inappropriate adduction of the vocal cords, it may also be caused by inappropriate movement of the pharyngeal wall muscles.<sup>2</sup> It is considered to be an hysterical conversion reaction which may be responsive to a wide range of psychological therapies.<sup>3</sup> Although initial reports suggested that it was a discrete isolated disorder, we have recently described a number of patients in whom paradoxical vocal cord adduction co-existed with documented asthma, and in whom objective evidence of refractory symptomatic paradoxical vocal cord adduction persisted for over a decade in spite of a wide range of psychotherapeutic interventions.<sup>4</sup> This present report further extends the spectrum of pulmonary disorders in which paradoxical vocal cord adduction may be found.

Many patients with cystic fibrosis have symptoms of wheeze and shortness of breath with a positive response to inhaled bronchodilators which may vary with pulmonary exacerbations of the disease.<sup>5</sup> To our knowledge there have been no previous reports of paradoxical vocal cord adduction in patients with cystic fibrosis. One of the puzzling psychodynamic features of this disease is its virtually exclusive occurrence in young women, the majority of whom have been involved in health care-related occupations. Our patient, like many individuals with cystic fibrosis, had been introduced to the hospital milieu from an early age and had many hospital admissions. Interestingly, the patient reported here had been heavily involved in life-saving courses, had spent considerable time as a beach lifeguard, and had expressed a desire to be a medical laboratory technician. This functional disorder is obviously rare, but it is clear that it could be easily overlooked in patients with cystic fibrosis who may present with various disorders of the upper and lower airways including recurrent respiratory infections, asthma, oropharyngeal candidiasis, and dysphonia due to inhaled steroid. We believe that the diagnosis of paradoxical vocal cord adduction should be considered in patients with cystic fibrosis in whom atypical chest symptoms, hoarseness, or stridor persist, and we suggest that appropriate physiological and endoscopic assessments are carried out.

- 1 Goldman J, Muers M. Vocal cord dysfunction. *Thorax* 1991; 46:401-4.
- 2 Appleblatt NH, Baker SR. Functional upper airways obstruction, a new syndrome. *Arch Otolaryngol* 1981;107: 305-7.
- 3 Martin RJ, Blager FB, Gay ML, Wood RP. Paradoxical vocal cord motion in presumed asthmatics. *Semin Respir Med* 1987;8:332-7.
- 4 Hayes JP, Nolan MT, Brennan N, FitzGerald MX. Three cases of paradoxical vocal cord adduction followed up over a ten year period. *Chest* 1993;104:678-80.
- 5 Hordvik NL, Konig P, Morris D, Kreutz C, Barbero GJ. A longitudinal study of bronchodilator responsiveness in cystic fibrosis. *Am Rev Respir Dis* 1985;131:889-93.