

Case reports

Management of respiratory failure with ventilation via intranasal stents in cystic fibrosis

F P Edenborough, M Wildman, D W Morgan

Abstract

The case history is presented of a patient with acute respiratory failure complicated by nasal obstruction resulting in intolerance of nasal ventilation. Urgent insertion of nasal stents permitted restoration of ventilation with resolution of breathlessness and stabilisation of arterial blood gases.

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Keywords: cystic fibrosis; respiratory failure; nasal stents; nasal intermittent positive pressure ventilation

Type II respiratory failure is common in patients with cystic fibrosis with advanced disease and may respond to long term nasal ventilation. We report a case of acute respiratory failure complicated by nasal obstruction resulting in intolerance of nasal ventilation which required urgent insertion of nasal stents.

Case history

The 28 year old man with cystic fibrosis had diabetes mellitus, mild liver disease, aminoglycoside induced renal tubulopathy, sputum colonised with *Pseudomonas aeruginosa*, and type I (hypoxic) respiratory failure requiring long term oxygen therapy. His spirometric test results when well gave forced expiratory volume in one second (FEV₁)/forced vital capacity (FVC) of 0.95/2.67 l with stable capillary blood gases as in table 1 (column A).

He was admitted with an FEV₁/FVC of 0.55/1.80 l and an oxygen saturation of 84% breathing room air. His nebulised medication included DNase (Pulmozyme®), β_2 agonists, and colistin sulphate with inhaled fluticasone. He received supplemental nasogastric feeding and required Creon 8000, cisapride, ranitidine, metoclopramide, and Humulin insulin.

Investigations revealed hypokalaemia (K^+ = 2.7 (normal range 3.5-5.3) mmol/l), hypomagne-

saemia (Mg^{2+} = 0.68 (normal range 0.7-1.0) mmol/l), and neutrophilia of 11 700/ml. A full blood count and renal and liver function were otherwise normal. Treatment with intravenous aztreonam and tobramycin and K^+ and Mg^{2+} supplementation was commenced and he initially improved, but after 14 days his antibiotics were changed to Tazocin, colistin sulphate, and ciprofloxacin for extended cover of a resistant *Pseudomonas* and metronidazole for possible anaerobic infection.

He continued to improve but on the 40th day of admission he again deteriorated with a fever of 39.6°C with arterial blood gas tensions as shown in table 1 (column B). Nasal intermittent positive pressure ventilation (NIPPV) was commenced at settings of 3/12/12 on 4 l oxygen (expiratory positive airway pressure (EPAP) 3 cm H₂O/inspiratory positive airway pressure (IPAP) 12 cm H₂O/default rate = 12 breaths/min). He deteriorated overnight (table 1 (column C)) and was treated with intravenous hydrocortisone, increased ventilation (3/14/12 on 6 l oxygen), and physiotherapy for presumed sputum retention. The antibiotics were changed to ceftazidime, tobramycin, and Tazocin and by the following morning he had improved (table 1 (column D)). The following night his nose blocked and, despite fluticasone 50 µg nasal spray, ephedrine 0.5% drops, and saline washes, he was unable to tolerate NIPPV either nasally or via a full face mask. Despite maximal medical treatment including intensive physiotherapy and maximal oxygen by mask his gases deteriorated (table 1 (column E)) and he began to tire. The changes in blood gas tensions are shown in fig 1.

Nasendoscopy performed to determine the cause of the nasal blockage revealed minimal crusting and debris but markedly oedematous nasal mucosa unresponsive to 1:80 000 adrenaline and 5% cocaine spray. An attempt to insert size 6 nasopharyngeal tubes failed due

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Table 1 Blood gas tensions during nasal ventilation in relation to placement of stents

	A	B(-3)	C(-2)	D(-1)	E(0)	F(+1)	G(+1)	H(+4)	I(+6)	J(+7)	K(+17)	L(+20)
NIPPV Settings	No	No	Yes	Yes	No	Yes	Yes	Yes	Yes	Yes	Yes	No
Fio ₂ (l)	—	—	3/9/12	3/11/12	3/11/12	3/11/12	3/13/12	3/13/15	3/13/15	3/13/15	3/13/15	—
Air	4	4	4	4	6	8	12	10	8	8	4	4
H ⁺	7.5	41.0	33.0	33.1	30.7	40.1	32.6	35.1	36.2	35.5	42.6	44.2
CO ₂	5.9	9.3	10.0	6.8	6.7	7.6	7.5	7.1	7.1	7.6	8.2	7.7
O ₂	7.5	6.7	7.3	8.3	5.0	6.4	9.6	8.7	9.6	18.2	17.2	11.2
HCO ₃	33.5	40.8	55.1	37.7	40.5	34.8	42.0	37.7	36.4	39.8	34.7	31.5
Sao ₂ (%)	92	84	91	94	75	82	95	93	95	99	94	—

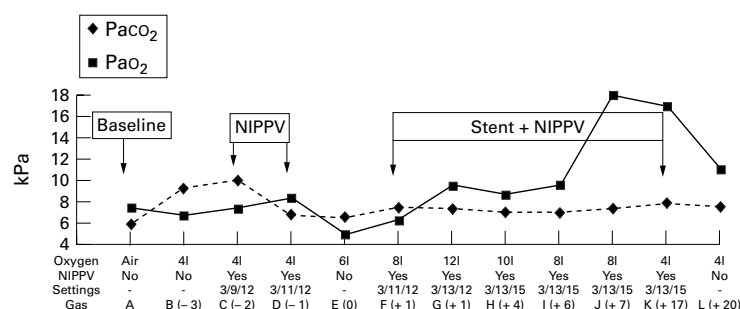


Figure 1 Blood gas tensions during nasal ventilation in relation to placement of stents.

to the extent of the obstruction airway. Portex Blue Line size 4.5 siliconised endotracheal tubes (internal diameter 4.5 mm, external diameter 6.5 mm, Portex Ltd, Hythe, Kent, UK) were therefore placed under direct vision using a nasendoscope to lie in the postnasal space on each side, trimmed at approximately 14 cm with 2 cm projecting externally allowing the tubes to be sutured together anterior to the columella (fig 2A and B). Nasal toilet was then performed with saline douches. Advancing the tubes into the nasopharynx (fig 2C and D) by extending the patient's neck enabled pulmonary toilet via a size 10 suction catheter after which the tubes were withdrawn back into the postnasal space to avoid excessive gagging. After two hours on NIPPV set at 3/11/12 with 8 l oxygen the gases were as shown in table 1 (column F) and fig 1, improving overnight

(table 1 (column G)) on increased ventilation (3/14/12 and 12 litres oxygen).

Continued maximal medical therapy including physiotherapy five times daily resulted in improved gas tensions despite reducing inspired oxygen. Nasogastric feeding was recommenced via a size 8 Portex infant feeding tube passed through the right sided stent. The left tube was removed on day 18 and by day 20, when no longer reliant on overnight ventilation (table 1 (column L)) the second stent was removed. Fluticasone 50 µg and xylometazoline 0.1% nasal sprays were commenced but the patient reported no nasal stuffiness or blockage and was later able to recommence NIPPV.

No organism was found to account for his pyrexial illness and deterioration. Multiple central and peripheral blood cultures, urine and stool cultures were negative. Tests for *Legionella* and respiratory viruses including CMV and EBV were also negative. His *Pseudomonas* remained sensitive to tobramycin and colistin sulphate throughout with intermittent sensitivity to Tazocin. At the time of writing he remains in hospital on intravenous antibiotics and on the active transplant list.

Discussion

Nasal polyposis and chronic rhinosinusitis is not uncommon in cystic fibrosis.¹ Nasal ventilation has previously been described in cystic

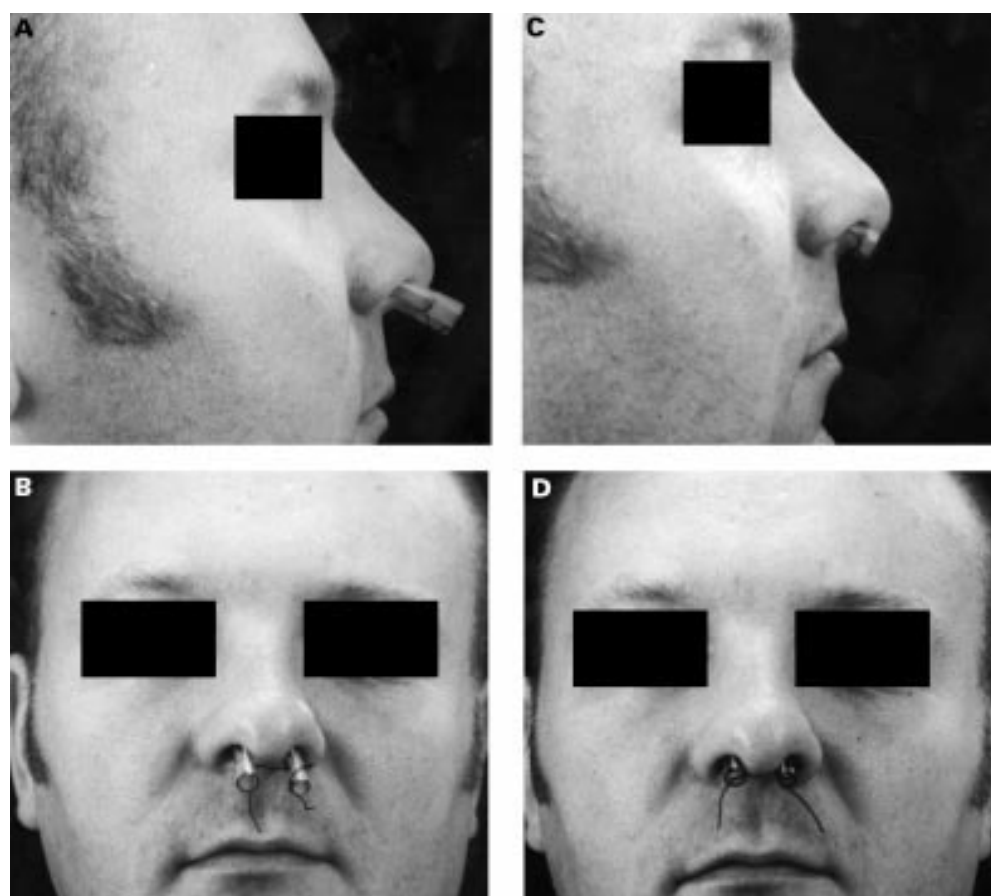


Figure 2 Nasal stents projecting (A) and (B) 2 cm beyond the nares over which the nasal mask fitted comfortably and (C) and (D) pushed into the nasopharynx to facilitate passage of a suction catheter and nasogastric tube.

fibrosis² and, in this centre,³ is used to treat type II respiratory failure in patients awaiting transplantation. Despite routinely prescribed nasal steroids, dryness and crusting is common though recently introduced custom designed warm air humidifiers such as HumidAire (Res-Med (UK) Ltd, Abingdon, Oxon, UK) appear to lessen the incidence of nasal problems.

Alternative patient interfaces with NIPPV are available including full face masks, unsuccessful in this case, and mouth only or nose and mouthpieces which have been used extensively elsewhere⁴ but which were not available in this case. Standard nasopharyngeal airways were found to be too soft to bypass the obstruction and hence an alternative was sought. Silicone tubes have been used as stents following surgery for acquired choanal atresia⁵ in infants and rarely in adults.⁶ Endotracheal tubes have similarly been used in infants⁷ but we are unaware of the use of this technique acutely in adults.

In this case, having been unable to oxygenate the patient, urgent insertion of nasal stents permitted restoration of NIPPV which successfully treated the acute hypoxia and improved hypercapnia over time. The procedure was well tolerated; nasal complications such as colu-

mellar or septal ulceration, nasal cavity or sinus infection were not seen⁸ and the nares remained patent after removal of the tubes. We believe the use of urgently inserted nasal stents was life saving in this case.

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Recurrent laryngeal nerve palsy associated with mediastinal amyloidosis

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Abstract

Amyloidosis affecting peripheral nerves causing isolated nerve palsies is uncommon. Localised amyloidosis occurs less frequently than the reactive or immune related systemic forms, and mediastinal localisation is virtually unknown. We present a case of recurrent laryngeal nerve palsy associated with mediastinal AL amyloidosis in a middle aged man.

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Keywords: amyloidosis; neuropathy; mediastinum

Case report

A 64 year old white retired bricklayer presented with a three week history of hoarseness and a five week history of dysphagia for solids. Apart from his hoarseness, physical examination was non-contributory. Chest radiography showed a grossly widened mediastinum. Despite a normal Kveim test and bronchial and transbronchial biopsy specimens, he had an unproven diagnosis of sarcoidosis based on a chest radiograph performed 17 years previously showing bilateral hilar lymphadenopathy and bilateral

upper lobar nodular shadowing. Computed tomographic scanning showed a large calcified mass in his superior mediastinum surrounding the trachea, the aortic arch, compressing the left brachiocephalic vein, and associated with bilateral calcified upper lobe masses (fig 1). A barium swallow showed a small upper oesophageal posterior diverticulum but no

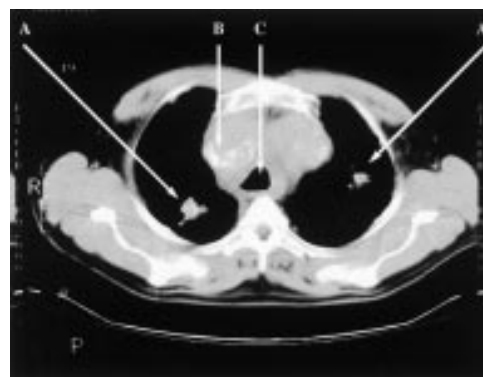


Figure 1 Computed tomographic scan showing mediastinal "amyloidoma" around the trachea. (A) Upper lobe amyloid deposits; (B) amyloidoma; (C) trachea.

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hold up of barium. His erythrocyte sedimentation rate (ESR) was 30 mm/h and lactate dehydrogenase (LDH) levels were 1030 U/l, but his serum angiotensin converting enzyme level was only mildly raised and serum calcium and phosphate levels were normal. Fiberoptic laryngoscopy revealed a paralysed left vocal cord. A right sided thorascopic biopsy of the mediastinal mass stained positively for amyloid with Congo Red and displayed apple-green birefringence with high intensity cross polarised light microscopy. It did not stain with antiserum against either serum amyloid A or immunoglobulin light chains, a result completely consistent with a diagnosis of AL amyloidosis. An associated inflammatory cell reaction consisted of small lymphocytes with only occasional typical plasma cells. No plasma cell dyscrasia or lymphoproliferative disorder was seen histologically. Bence Jones proteinuria was not demonstrated, but serum monoclonal immunoglobulin G lambda (λ) light chain paraprotein was present. A serum amyloid P-component scan, prostatic and bronchial biopsy specimens were all negative, suggesting that the mediastinal amyloidosis was localised. Bone marrow biopsy specimens were also normal. Over the next two months his dysphagia resolved spontaneously. His hoarseness was improved greatly by teflon injection of the left vocal cord and speech therapy. One year after the biopsy there is no clinical or radiological progression of the disease and no further treatment is considered at this stage.

Discussion

Amyloid is described as an homogeneous, proteinaceous, eosinophilic fibrillar deposit in various tissues. However, x ray diffraction accurately identifies the amyloid fibril by its unique β -pleated sheet conformation.¹ Diagnostic characteristics are: (1) apple-green birefringence of Congo Red stained tissue under polarised light, (2) bright yellow-green fluorescence in thioflavine stained preparations, and (3) ultrastructural demonstration of 7–10 nm non-branching fibrils of indeterminate length.^{2–3} Primary immunoglobulin light chain (AL) amyloid occurs in its localised form in extramedullary plasmacytomas of the upper airways and gastrointestinal tract. However, localisation of amyloid to the mediastinum is rare, with only two cases reported as the AL

type⁴ and one as the reactive (AA) form.⁵ Although calcific changes were evident on the chest radiograph, both bronchial and trans-bronchial biopsy specimens were normal. No histological diagnosis has been obtained of the upper lobe lesions but they are also considered to be amyloid deposits.

It is possible that the earlier chest radiographic findings diagnosed as sarcoidosis could represent early amyloid deposits, although the radiographic changes are typical for sarcoidosis.

Apart from carpal tunnel syndrome associated with β_2 -microglobulin deposition in haemodialysis patients, amyloid rarely affects the peripheral nervous system and is usually of the λ type, being of monoclonal origin in 85% of cases.⁶ Familial patterns of amyloid mixed peripheral polyneuropathy have been described, commonly associated with variant plasma transthyretin.⁷ This case of recurrent laryngeal nerve palsy associated with amyloidosis may be due to perineural infiltration or compression. Infiltration of the vagus nerve itself with amyloid may account for both the vocal cord paralysis and oesophageal dysmotility.

The mediastinal "amyloidoma" may represent an underlying lymphoplasmacytoma or a subtle plasma cell dyscrasia such as a Castleman's disease tumour of the solitary plasma cell variety, although this patient had no constitutional symptoms or signs of a POEMS syndrome. No steroids or cytotoxic treatment have been considered. Technically challenging surgical resection of this amyloid mass is best reserved for severe compressive and obstructive symptoms of the vascular, upper airway, or oesophageal lumen. Alternatively, intraluminal stenting may be considered.

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