

Editorial

Assisted ventilation for chronic neuromuscular disorders

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The provision of long term ventilatory support to patients with chronic neuromuscular disorders is a major logistic, technical, and clinical challenge. In this issue of *Thorax* Baydur and colleagues share their experiences with ventilatory support in this context.¹ Much of their experience reflects the advances in technology that have evolved over the last six decades. Their initial experience was with negative pressure ventilation and later with positive pressure ventilation. Positive pressure ventilation has evolved from an invasive interface (the tracheostomy) to, in many situations, a non-invasive interface. Although negative pressure ventilation served their early patients well in the post-poliomyelitis epidemic, their experience with non-invasive ventilation—mouth or nasal intermittent positive pressure ventilation (MNIPPV)—indicates that, in both poliomyelitis and Duchenne's muscular dystrophy, MNIPPV is more effective.

The slow decline in forced vital capacity (FVC) over 24 years in the poliomyelitis group receiving negative pressure ventilation is remarkable. In the 25 patients studied the FVC declined by an average of 5 ml per year (0.1% predicted per year) from a mean of 800 ml to a mean of 730 ml. In the albeit shorter time interval of three years their patients with poliomyelitis receiving MNIPPV had an actual increase in vital capacity from a mean of 1134 ml to 1219 ml, a mean increase of 90 ml/year. Even if their lung function were to decline at the rate of those on negative pressure ventilation, they may not reach the point of ventilatory muscle weakness that had previously been associated with the institution of invasive ventilation (tracheostomy). Bach *et al*² found that long term ventilation of patients with a history of poliomyelitis using mouth IPPV resulted in a decline in FVC of 19 ml/year. The patients in this study had slightly lower FVC values to start with which may explain the more rapid decline in lung function, and the patients were followed for 15 years rather than 24 years as in the study by Baydur *et al*.

In patients with Duchenne's muscular dystrophy the decline in lung function was more rapid. At the onset of mechanical ventilation the FVC values of the patients were strikingly lower than those with poliomyelitis with a mean of 600 ml falling to 533 ml (60 ml/year) over the two years that they received treatment with MNIPPV. The decline in FVC in these patients is similar to that reported previously.³ This decline in lung function from an already low initial value resulted in a tracheostomy in 10 of the 15 patients, in most cases because of their inability to ventilate adequately with non-invasive techniques. Patients with negligible vital capacity and without bulbar involvement have been managed with non-invasive ventilation.⁴ The experience of Baydur *et al* is similar to that of Leger *et al*⁵ who also found that a tracheostomy became necessary in many of their patients with Duchenne's muscular dystrophy.

Can we manage such patients with profound ventilatory muscle weakness completely non-invasively in a routine fashion? The challenge of managing these patients 24 hours a day with non-invasive devices is significant. Patient motivation and the availability of an infrastructure to provide the appropriate interfaces with the ventilator appear critical. The use of interfaces applied to the mouth appear more important as weakness progresses.⁴ Failure to achieve control of ventilation often precipitates tracheostomy to secure breathing. Future improvements in technology and increased familiarity with the available techniques should facilitate the application of non-invasive ventilation in these patients.

The concept of initiating ventilation earlier in an attempt to prevent the inevitable decline in lung function, particularly in patients with Duchenne's muscular dystrophy, is laudable. Unfortunately the only randomised controlled clinical trial to assess the effectiveness of early intervention in Duchenne's muscular dystrophy had a negative result.⁶ The idea that intermittently resting overstimulated muscle units in patients with poliomyelitis or preventing muscle injury induced by overexertion in patients with muscular dystrophy^{7,8} is a sound one. The fact that the respiratory muscles may also be injured lends relevance to this idea.⁹ The finding that muscle contraction itself can result in muscle injury and subsequent cell necrosis in patients with muscular dystrophy supports the notion that intermittent rest may be beneficial. Complete rest will, of course, lead to muscle atrophy. A re-examination of the potential effects of rest on ventilatory muscle weakness in this condition will probably depend on improved usage and techniques of ventilatory assistance. At present, however, ventilation—both invasive and non-invasive—allows the prolongation of a meaningful life in patients with Duchenne's muscular dystrophy and, until a cure is found for this disease, the next obstacle confronting these individuals will be failure of the heart as a result of cardiomyopathy. In general, however, the paper by Baydur and colleagues serves to highlight the very viable option of assisted ventilation, invasive or non-invasive, in patients with chronic neuromuscular disease and chest wall disorders.¹⁰ This group of patients can clearly benefit from this intervention.

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- 1 Baydur A, Layne E, Aral H, *et al*. Long term non-invasive ventilation in the community for patients with musculoskeletal disorders: 46 year experience and review. *Thorax* 2000;55:4-11.
- 2 Bach JR, Alba AS, Bohatuk G, *et al*. Mouth intermittent positive pressure ventilation in the management of postpolio respiratory insufficiency. *Chest* 1987;91:859-64.
- 3 Baydur A, Gilgoff I, Prentice W, *et al*. Decline in respiratory function and experience with long-term assisted ventilation in advanced Duchenne's muscular dystrophy. *Chest* 1990;97:884-9.
- 4 Bach JR. The prevention of ventilatory failure due to inadequate pump function. *Respir Care* 1997;42:403-13.

- 5 Leger P, Bedican JM, Cornette A, *et al.* Nasal intermittent positive pressure ventilation. Long-term follow-up in patients with severe chronic respiratory insufficiency. *Chest* 1994;105:100-5.
- 6 Raphael JC, Chemet S, Chastang C, *et al.* Randomized trial of preventive nasal ventilation in Duchenne's muscular dystrophy. *Lancet* 1994;343:1600-4.
- 7 Johnson EW, Barddon R. Over-work weakness in facioscapulohumeral muscular dystrophy. *Arch Phys Med Rehabil* 1971;52:333-4.
- 8 Petrof BJ, Shrager JB, Stedman HH, *et al.* Dystrophin protects the sarcolemma from stresses developed during muscle contraction. *Proc Natl Acad Sci USA* 1993;90:3710-4.
- 9 Road JD, Jiang TX. Determinants of diaphragmatic injury. *Mol Cell Biochem* 1998;179:81-6.
- 10 Chailleux E, Fauroux B, Binet F, *et al.* Predictors of survival in patients receiving domiciliary oxygen therapy or mechanical ventilation. *Chest* 1996;109:741-9.

Annual report

Annual report October 1998 to September 1999

1999 has been yet another successful year for *Thorax* with an improved impact factor (2.861, ISI Statistics, 1999) of which all who contribute to the journal can be proud.¹ These new figures reflect the fact that we are now receiving work of the highest quality. Our initial policy aim of attracting high quality submissions is now taking effect, and we still believe that the journal can continue to improve and provide a competitive international forum for the best research in respiratory medicine.

Thorax has continued its development during the last year. The number of submissions remains high; out of 812

Table 1 Articles submitted by category

	1995	1996	1997	1998	1999
Original papers	493	415	411	398	453
Case reports	252	247	212	211	149
Short papers	48	51	40	31	36
Editorials/reviews	38	38	53	81	57
Review series	18	31	26	13	19
Supplement articles	16	41	29	17	16
Case report commentaries	7	11	7	7	0
Rapid communications	—	5	5	7	3
Total	872	839	783	765	733

Table 2 Geographical distribution of submissions

	1995	1996	1997	1998	1999
UK	291	320	303	265	326
Western Europe	266	264	236	242	230
Spain	46	42	45	44	37
The Netherlands	51	44	49	41	40
Italy	38	48	19	38	40
France	27	28	24	27	26
Turkey	13	14	22	20	17
Israel	20	17	15	10	13
Eire	10	8	7	5	2
Switzerland	10	13	9	5	5
Germany	16	26	23	20	29
Belgium	17	12	11	10	10
Greece	7	7	6	15	6
Austria	7	5	5	3	4
Portugal	3	0	0	2	0
Malta	1	0	0	2	1
USA and Canada	101	71	84	76	81
Japan	65	53	64	46	54
Australasia	38	31	60	48	38
Scandinavia	36	32	39	36	38
Asia	34	26	26	35	30
Eastern Europe	7	10	3	6	4
South America	14	10	3	3	1
Africa	7	4	4	3	3
Middle East	13	5	9	5	7

Table 3 Median time to first decision (days)

	1995/6	1996/7	1997/8	1998/9
Full papers	74	44.5	56	48.5
Short papers	82	38	24	11.5

Table 4 Decision of papers submitted 1998/9 as at 30 September 1999

	Case reports	Papers	Short papers
Total	149	453	36
Rejected	129 (87%)	281 (62%)	24 (67%)
Accepted	5 (3%)	51 (11%)	4 (11%)
Pending	15 (10%)	121 (27%)	7 (19%)
Withdrawn			1 (3%)

total submissions (including letters), 453 were original papers, the highest figure for five years (table 1), and in September we had the highest number submitted in a month since we became editors. As can be seen from table 2, the papers come from all over the world with little noticeable difference from year to year in numbers of submissions, and the UK remaining at the head of the table. We encourage contributions from all areas of the world.

Once again we have improved the speed of our review system (table 3) and we aim to keep authors informed as much as possible of the progress of their manuscript. We have successfully reached our objective of a three month period between acceptance and publication of original articles. We also aim to speed up time to publication and will be introducing the electronic despatch of proofs in the very near future.

Table 4 shows the rates of acceptance which, though showing only 11% of papers accepted each year (this will be higher when pending papers have reached their conclusion), means that only those of the highest quality appear in the journal and there is no large backlog of original papers awaiting publication at any one time. Editorial policy to accept and therefore publish fewer case reports has been reflected in the reduction in number submitted (table 1).

The past year has seen the successful publication of important guidelines on smoking cessation² and the management of diffuse parenchymal lung disease.³ While review series on passive smoking and genetics drew to a close, we saw the arrival of informative series on rare diseases⁴ and the interface between primary and secondary care in the management of respiratory disease,⁵ with a

series on the paediatric origins of adult lung disease due to appear in the New Year. 1999 saw the publication of the fifth Year in Review,⁶ another useful resource for continuing education.

February 1999 saw the arrival of *eThorax*⁷ which has proved to be a popular research tool with many highly developed new features. Access is free to subscribers to the journal and a web only subscription is now available. The site includes full text, fully searchable archive, articles collected by topic, customised email alerts, cite track, direct access to Medline, and links to a wide range of other journals.

We would like to thank all who work behind the scenes at *Thorax* for their continuing hard work, especially the excellent support of Liz Stockman and Rachel Harvey, and all who have helped with the development and maintenance of the website. We would also like to thank the Year in Review editors and contributors, and the editors of our review series. *Thorax* is also grateful for all the hard work of the associate editors and advisory board, and the essential contribution of the referees who are acknowledged on page 94.

Many thanks to all who have contributed to the journal in the past year, and we hope *Thorax* has managed to inform and entertain.

J BRITTON
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Editorial Assistants

- 1 Britton JR, Knox AJ. Impact factors for 1998. *Thorax* 1999;54:960.
- 2 Smoking cessation guidelines and their cost effectiveness. *Thorax* 1998; 53(Suppl 5):S1-18, S1-37.
- 3 British Thoracic Society, Standards of Care Committee. The diagnosis, assessment and treatment of diffuse parenchymal lung disease in adults. *Thorax* 1999;54(Suppl 1):S1-28.
- 4 Tattersfield AE, du Bois RM. Rare diseases: introduction. *Thorax* 1999;54: 253.
- 5 Holmes WF, Macfarlane J. Issues at the interface between primary and secondary care in the management of common respiratory disease: introduction. *Thorax* 1999;54:538-9.
- 6 Mitchell DM, Woodcock A, eds. 1999 Year in Review: selected topics in respiratory medicine from 1998. *Thorax* 1999;54(Suppl 2):S1-62.
- 7 Orme R. *Thorax* with extra "e". *Thorax* 1999;54:189.