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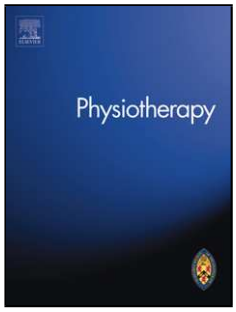

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Airway clearance techniques used by people with cystic fibrosis in the UK

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Abstract

Objectives To describe the current use of airway clearance techniques among people with cystic fibrosis (CF) in the UK, and the baseline characteristics for users of different airway clearance techniques.

Design Analysis of the UK CF Registry 2011 data.

Setting and participants All people with CF in the UK aged ≥11 years (*n*=6372).

Results Of the 6372 people on the UK CF registry in 2011, 89% used airway clearance techniques. The most commonly used primary techniques were forced expiratory techniques (28%) and oscillating positive expiratory pressure (PEP) (23%). Postural drainage and high-frequency chest wall oscillation were used by 4% and 1% of people with CF, respectively. The male:female ratio of individuals who used exercise as their primary airway clearance technique was 2:1, compared with 1:1 for other techniques. Individuals with more severe lung disease tended to use devices such as non-invasive ventilation or high-frequency chest wall oscillation.

Conclusions Forced expiratory techniques and oscillating PEP are the most common airway clearance techniques used by people with CF in the UK, and postural drainage and high-frequency chest wall oscillation are the least common techniques. This is significant in terms of planning airway clearance technique trials, where postural drainage has been used traditionally as the comparator. The use of airway clearance techniques varies between countries, but the reasons for these differences are unknown.

Keywords: Cystic fibrosis; Physiotherapy; Registries; Epidemiology

Introduction

Lung disease in people with cystic fibrosis (CF) is caused by retained airway secretions, inflammation and infection [1]. Chest physiotherapy to remove airway secretions is recommended by various CF organisations as a vital treatment for people with CF [2,3].

Many different airway clearance techniques have been developed and modified to optimise outcomes in CF [4]. Despite various studies and meta-analyses (including five Cochrane systematic reviews), the best airway clearance technique for people with CF has yet to be identified [5]. As such, it is recommended that airway clearance techniques should be tailored according to individual needs, lifestyle and symptoms [2,3]. Most people with CF have their own preferences for a particular airway clearance technique, as shown by the drop-outs in a recent randomised controlled trial of long-term airway clearance techniques [6].

As such, the airway clearance techniques used by people with CF vary between individuals attending the same CF centre, between CF centres in the same country, and between countries. An understanding of this variation would be useful in terms of planning future airway clearance technique trials, but the airway clearance techniques used in the UK have not been explored systematically. This paper aims to describe the airway clearance techniques used by adolescents and adults with CF in the UK, to explore the baseline characteristics of people with CF using different airway clearance techniques, and to investigate usage patterns of airway clearance techniques by severity of lung disease.

<A>Methods

A cross-sectional analysis of all available data from the UK CF registry was undertaken for patients aged ≥11 years in 2011. The study had no exclusion criteria. Only data from people with CF aged ≥11 years were included because airway clearance techniques suitable for younger children differ from those used by adolescents and adults [2]. Also, the inclusion of data from younger children

would move the focus towards techniques that are chosen as much for the needs of parents and families, who are more involved at that stage, as the needs of the person with CF.

<B>*Data*

The following data were obtained from the registry:

- demographics – age, sex;
- spirometry – forced expiratory volume in 1 second (FEV_1) (in % predicted);
- annual total intravenous (IV) antibiotic-days (in number of days);
- use of mucolytic/osmotic – Dornase Alpha (DNase), nebulised hypertonic sodium chloride; and
- primary airway clearance – recorded within the UK CF registry as postural drainage, forced expiratory techniques (e.g. active cycle of breathing techniques), positive expiratory pressure (PEP), oscillating PEP (e.g. Flutter valve and Acapella), high-frequency chest wall oscillation (e.g. vest), other (usually non-invasive ventilation), exercise and none specified.

Data were collected during the annual review of people with CF between January and December 2011. Of note, the CF Trust made minor modifications to the airway clearance technique categories in 2013, and the 'forced expiratory techniques' category was removed as forced expiratory techniques are now recognised to be an integral part of all airway clearance techniques in the UK. This removal was made following consultation with specialist physiotherapists working within CF care who were concerned that this 'catch all' heading did not allow differentiation between very different airway clearance techniques, such as autogenic drainage and active cycle of breathing techniques.

Analyses were restricted to primary airway clearance techniques due to missing data for secondary airway clearance techniques (no data were available for 51% of people with CF).

<B>*Statistical analysis*

Excel 2010 (Microsoft Corp., Redmond, WA, USA) spreadsheets received from the UK CF Registry were converted into SPSS Version 20 (IBM Corp., Armonk, NY, USA) spreadsheets for data cleaning and analysis.

The distribution of primary airway clearance techniques was described. Descriptive analysis of baseline characteristics (demographics, spirometry, annual total IV antibiotic-days, use of mucolytic/osmotic) was obtained for each airway clearance technique.

Severity of lung disease was divided into severe (% predicted FEV_1 ≤40%), moderate (% predicted FEV_1 >40% to <70%) and mild (% predicted FEV_1 ≥70%). These divisions are used as the standard for epidemiological studies in CF [7], and are used by US and Canadian CF registries [8,9]. The robustness of these divisions for this dataset was tested by comparing the total IV antibiotic-days for each group using the Kruskal-Wallis test. Annual total IV antibiotic-days increased with the severity of lung disease (Kruskal-Wallis $P<0.0001$), suggesting that the internationally adopted divisions were applicable to this dataset.

<insert Figure 1 near here>

Chi-squared test was used to test associations between airway clearance techniques, use of mucolytic/osmotic and severity of lung disease. All statistical tests were two-sided, and $P<0.05$ was considered to indicate significance.

<A>**Results**

In total, 6372 people with CF aged ≥11 years were registered on the UK CF registry database in 2011. There were no missing data for age and sex, and minimal missing data (<5%) for FEV_1 and annual total IV antibiotic-days, as shown in Table 1. Missing data for use of DNase and nebulised hypertonic sodium chloride were difficult to ascertain as the data were collected by the UK CF Registry with a check-box, with no option for 'not using'. Therefore, if the check-box was not

selected (i.e. 'no data'), it could be that the respondent was not using that mucolytic/osmotic or it could represent missing data.

<insert Table 1 near here>

Fig. 2 shows that 89% of people with CF used airway clearance techniques in 2011. Forced expiratory techniques (including huff, active cycle of breathing techniques and autogenic drainage), used by 28% of people with CF, were the most common, followed by oscillating PEP (23%). Only 1% of people with CF in the UK used high-frequency chest wall oscillation as their primary airway clearance technique. Approximately 10% of people with CF did not use any airway clearance techniques.

<insert Fig. 2 near here>

Table A (see online supplementary material) shows the demographic data by airway clearance technique. People with CF who used exercise and those who did not use any airway clearance techniques appeared to have the mildest lung disease based on % predicted FEV_1 (median 78% and 78%, respectively) and annual IV antibiotic-days (median 3 days and 0 days, respectively). People with CF using high-frequency chest wall oscillation or 'other' techniques (usually non-invasive ventilation) appeared to have the most severe lung disease, with median % predicted FEV_1 of 63% and 65%, respectively. These two groups also had the highest annual IV antibiotic-days (median 26 days and 24 days, respectively). Further information about the distribution of continuous variables (age, FEV_1 and total IV antibiotic-days) is available in Fig. A (see online supplementary material).

People with severe lung disease were more likely to use high-frequency chest wall oscillation [relative risk (RR) 2.75, 95% confidence interval (CI) 1.39 to 5.41] and forced

expiratory techniques (RR 1.79, 95% CI 1.61 to 1.99) than people with mild lung disease. People with mild lung disease were more likely not to use any airway clearance techniques (RR 1.40, 95% CI 1.09 to 1.80), or to use exercise (RR 1.98, 95% CI 1.59 to 2.48) or PEP (RR 1.79, 95% CI 1.45 to 2.23) as their primary airway clearance technique compared with people with severe lung disease. The use of postural drainage and oscillating PEP was similar across severity categories. Table 2 shows the proportion of airway clearance techniques by severity of lung disease (Chi-square, $P<0.001$).

<insert Table 2 near here>

Table 3 shows the use of mucolytic/osmotic by severity of lung disease. People with severe lung disease were more likely to use DNase than people with mild lung disease (RR 1.65, 95% CI 1.55 to 1.75; overall Chi-square, $p<0.0001$). People with severe lung disease were more likely to use nebulised hypertonic sodium chloride than people with mild lung disease (RR 1.88, 95% CI 1.63 to 2.17; overall Chi-square, $p<0.0001$).

<insert Table 3 near here>

As shown in Table A (see online supplementary material), people with CF who did not use any airway clearance techniques were least likely to use DNase (29%, compared with the overall figure of 53%) and nebulised hypertonic sodium chloride (11%, compared with the overall figure of 19%). On the other hand, people with CF who used high-frequency chest wall oscillation or 'other' techniques (usually non-invasive ventilation) were most likely to use DNase (67% and 67%, respectively) and nebulised hypertonic sodium chloride (29% and 30%, respectively).

<A>Discussion

<B>Levels of airway clearance technique use in the UK

Instead of 'traditional' techniques such as postural drainage (used by 4% of people with CF), more people with CF used techniques that can be self-administered: forced expiratory techniques (28%), oscillating PEP (23%) and PEP (16%). This is not surprising given that the 'traditional' techniques are relatively cumbersome and time-consuming to perform [10]. Postural drainage has gone out of favour due to the time required to facilitate mucociliary clearance [11]. There are also concerns that the head-down tilt may exacerbate gastro-oesophageal reflux and dyspnoea [12].

Three of the four recent randomised controlled trials on airway clearance techniques used PEP or oscillating PEP in one of the comparator arms [13–15]. Given their level of use across different countries and the relative evidence of benefit, all future randomised control trials on airway clearance techniques should consider using PEP or oscillating PEP as a comparator.

<B>Variation by country

There is considerable between-county variation in airway clearance techniques (Table 4). For example, more than 50% of people with CF at the Buenos Aires CF centre in Argentina used postural drainage as their main airway clearance technique [16], compared with 4% in the UK. High-frequency chest wall oscillation is used by 37% of people with CF in the USA [17], but only 3% in Canada [18] and 1% in the UK. Exercise is the most common airway clearance technique in New Zealand (46% of adults) [19], compared with 16% in the UK. There is a strong preference for autogenic drainage in Ireland (46% of adults) [20], with other countries reporting autogenic drainage grouped with other breathing techniques (e.g. active cycle of breathing techniques) and figures ranging from 5% [18] to 33% [16].

Review of registry data for other countries revealed that airway clearance data for Australia, Brazil, France, Germany or the Netherlands were not included in the annual registry reports [21–25]. Airway clearance data were also omitted from the European Cystic Fibrosis Society registry report [26]. However, 2010 data were available for Belgium, and showed that 90% of people with

CF on the registry underwent regular chest physiotherapy; no data were available regarding the actual technique used [27].

<insert Table 4 near here>

In 2007, the Canadian Cystic Fibrosis Foundation created an Airway Clearance Working Group to collect airway clearance technique data from their CF clinics [18]. The results of this survey influenced the choice of airway clearance techniques used for the comparator and intervention arms in a recent multicentre randomised control trial [13]. The proportions of the different airway clearance techniques recorded for people with CF in the UK are broadly comparable with Canada, except for a bias towards PEP over oscillating PEP in Canada. This difference may be explained by several Canadian studies which found that PEP therapy is more effective than flutter devices (a form of oscillating PEP) [28] and postural drainage [29]. Interestingly, data from the 2011 Canadian registry report differed from this study: 34% of people with CF used PEP, 29% used percussion, 9% used other techniques, 8% used no airway clearance, 7% used autogenic drainage or 'breathing exercises', 7% used high-frequency chest wall oscillation and 6% used flutter devices [30]. The differences in the Canadian data over time may be due to the 4-year gap between data collection, or issues such as data input and definitions.

<B>Limitations in using registry data

Limitations in using registry data to assess use of airway clearance techniques by people with CF were demonstrated in the above descriptions of data from other countries. Many reports did not include any information about airway clearance, and some reports only included minimal data with no information about the technique used. Technique limitations were also displayed in this analysis of UK registry data because the airway clearance techniques available in 2011 were less well defined, with no clear option for active cycle of breathing techniques or autogenic drainage. Instead,

these different techniques were combined into a single category (forced expiratory techniques), which is likely to have affected the results of this registry analysis which found that forced expiratory techniques were the most common primary techniques for people with CF. This limitation has been addressed for UK registry data entered from 2013.

Other limitations are that CF registry data does not include information regarding adherence, opinions of people with CF, exact 'prescription' and health-related quality of life. This is evidenced by the low percentages of people with CF who reported that they did not use any airway clearance techniques: 10% in this UK study, 4% in the New Zealand data, 8% in the Canadian registry data and no other reports within the other data identified in this paper [16–18,20]. Given that previous studies have often identified poor adherence to airway clearance, it seems unlikely that this is accurate.

Data input may also be an issue as the accuracy of the data is highly dependent on the clear labelling of categories within the registry, and the knowledge of the person inputting the data or providing the data to input. Non-physiotherapists may struggle to define what the patient is doing, and which category this belongs to within a registry database.

It is likely that people with CF use more than one airway clearance technique. The authors were unable to derive meaningful conclusions from the 'secondary' airway clearance technique data because no data were available for 51% of the people with CF, while less than 1% of the people with CF were not using any secondary airway clearance techniques. Nonetheless, it is expected that the registry data will be more robust following modifications made since 2013, and the implementation of a National Tariff for Cystic Fibrosis (payment by results) which will use registry data.

The advantage of analysing the CF registry database is that a snapshot for all registered people with CF in the UK is available. As a result, this analysis has a larger sample size compared with previous analyses [16–18]. The UK CF registry database also has stringent quality control systems [31]. The small amount of missing data (<2% of data for 'primary' airway clearance

technique categories were missing) is testament to the high quality and comprehensive cover of the registry. The descriptive statistics provided in this paper are therefore representative of the main airway clearance techniques used by people with CF in the UK.

<B>Baseline characteristics

To the authors' knowledge, this is the first paper is to describe the baseline characteristics of people with CF using various airway clearance techniques. People with mild lung disease were most likely to use exercise alone as their primary airway clearance technique, while people with severe lung disease were most likely to use high-frequency chest wall oscillation and nebulised mucolytics/osmotics. These results indicate a good degree of individualisation of airway clearance techniques by CF specialist physiotherapists, with some techniques (e.g. high-frequency chest wall oscillation, non-invasive ventilation) being reserved for sicker patients. However, recent trials comparing high-frequency chest wall oscillation with PEP have raised uncertainty regarding the effectiveness of high-frequency chest wall oscillation for airway clearance [13,32]. It will be interesting to repeat a similar analysis in several years' time to determine if the pattern of airway clearance techniques changes with emerging evidence.

There was a 50%:50% split by sex for most airway clearance technique categories. However, the male:female ratio for people with CF using exercise as their primary airway clearance technique was almost 2:1. Given that the sex difference in habitual activities among people with CF is well described [33], this result suggests that patient preference also exerts a significant influence on the choice of airway clearance technique. In addition, people with CF who do not use any primary airway clearance technique had high % predicted FEV_1 (median 78%) and low total IV antibiotic-days (median 0), suggesting that these people may have felt well and therefore chose not to perform airway clearance.

<B>Patient preference

Patient preference is particularly pertinent in airway clearance because these are time-consuming, effortful and demanding activities to perform [6]. Adherence is crucial for an airway clearance technique to work optimally [4], and self-reported adherence correlates best with self-reported satisfaction with a particular airway clearance technique [10]. There is emerging evidence that giving people with CF a choice of airway clearance techniques may improve outcome [34]. Nonetheless, patient preference alone does not guarantee efficacy – in the 2010 US multicentre randomised control trial on airway clearance techniques (whereby participants were randomised to different airway clearance techniques without taking their preferences into account), the attrition rate from the high-frequency chest wall oscillation arm was significantly lower compared with the oscillating PEP and postural drainage arms [15]. 'Satisfaction with therapy' was an independent predictor of drop-out [15]. However, the decrease in forced expiratory flow 25–75% ($FEF_{25–75}$) in the high-frequency chest wall oscillation arm was significantly higher compared with the other two arms [15], suggesting a potential mismatch between objective and perceived efficacy [6].

This analysis found that approximately 15% of people with CF in the UK use exercise as their primary airway clearance technique. People with CF may perceive exercise to be a non-stigmatising activity performed by all healthy people, so may choose to use exercise alone as an airway clearance technique despite recommendations to the contrary [2,3]. Evidence suggests that although exercise helps increase ease of sputum expectoration, it may not actually increase solid content [35]. However, exercise has numerous other benefits and should be strongly encouraged as part of CF management and as an adjunctive therapy for airway clearance [36].

<B>Adherence

Other than the potential lack of efficacy of the preferred technique, another reason for the mismatch between personal preference and actual efficacy of the airway clearance techniques may simply be overestimation of adherence to certain airway clearance techniques. The lack of adherence data within registries is a limitation. However, issues have also been raised around how adherence could

be reported if registries were to collect this data. Electronic data capture (EDC) has demonstrated that self-report among people with CF is notoriously unreliable. For example, people with CF reported a median adherence of 80% to nebulised treatment, but objective median adherence was only 36% [37]. Airway clearance has historically been invisible, with only self-report or crude time loggers to guide visualisation of adherence. Making airway clearance and physical activity visible with EDC will be an important step towards more meaningful analysis of the results of future comparative trials of airway clearance techniques. Physical activity can be quantified objectively with an accelerometer [38], and the use of PEP and oscillating PEP may be measured with electronic chip card recording.

<A>Conclusion

This UK CF registry analysis found that many people with CF do not use any airway clearance techniques, or use exercise as their primary airway clearance technique despite the fact that this is not recommended [2,3]. Forced expiratory techniques and airway clearance devices (PEP and oscillating PEP) were found to be the most common primary airway clearance techniques used by people with CF in the UK, while postural drainage and high-frequency chest wall oscillation are the least common techniques (used by 4% and 1%, respectively). This is a significant finding in terms of planning airway clearance technique trials where postural drainage has been used traditionally as the 'gold standard' comparator. Continuation of this practice would risk the recruitment of sufficient participants to a trial, and calls into question the applicability of the results to a population that does not tend to use these techniques.

This work demonstrates variation in the use of different airway clearance techniques between countries. It does not explain whether these differences are influenced by technique accessibility, clinician cultural or knowledge differences, patient cultural or knowledge differences, overall differences in emphasis of different treatments or other factors. This poses an important

research question to identify the factors that cause variation in the use of different airway clearance techniques between countries.

Acknowledgement

Data were obtained from the UK CF Registry, Cystic Fibrosis Trust, 2013. Part of the analysis in this paper will be presented as a poster in the 2014 European Cystic Fibrosis Society conference, and will also be published as an abstract in the 2014 Supplement 1 issue of the Journal of Cystic Fibrosis (titled 'Airway clearance techniques used by people with cystic fibrosis in the UK').

Ethical approval: None required.

Conflicts of interest: None declared.

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Fig. 1. Box-and-whisker plots of total intravenous (IV) antibiotic-days by severity of lung disease. FEV_1, forced expiratory volume in 1 second.

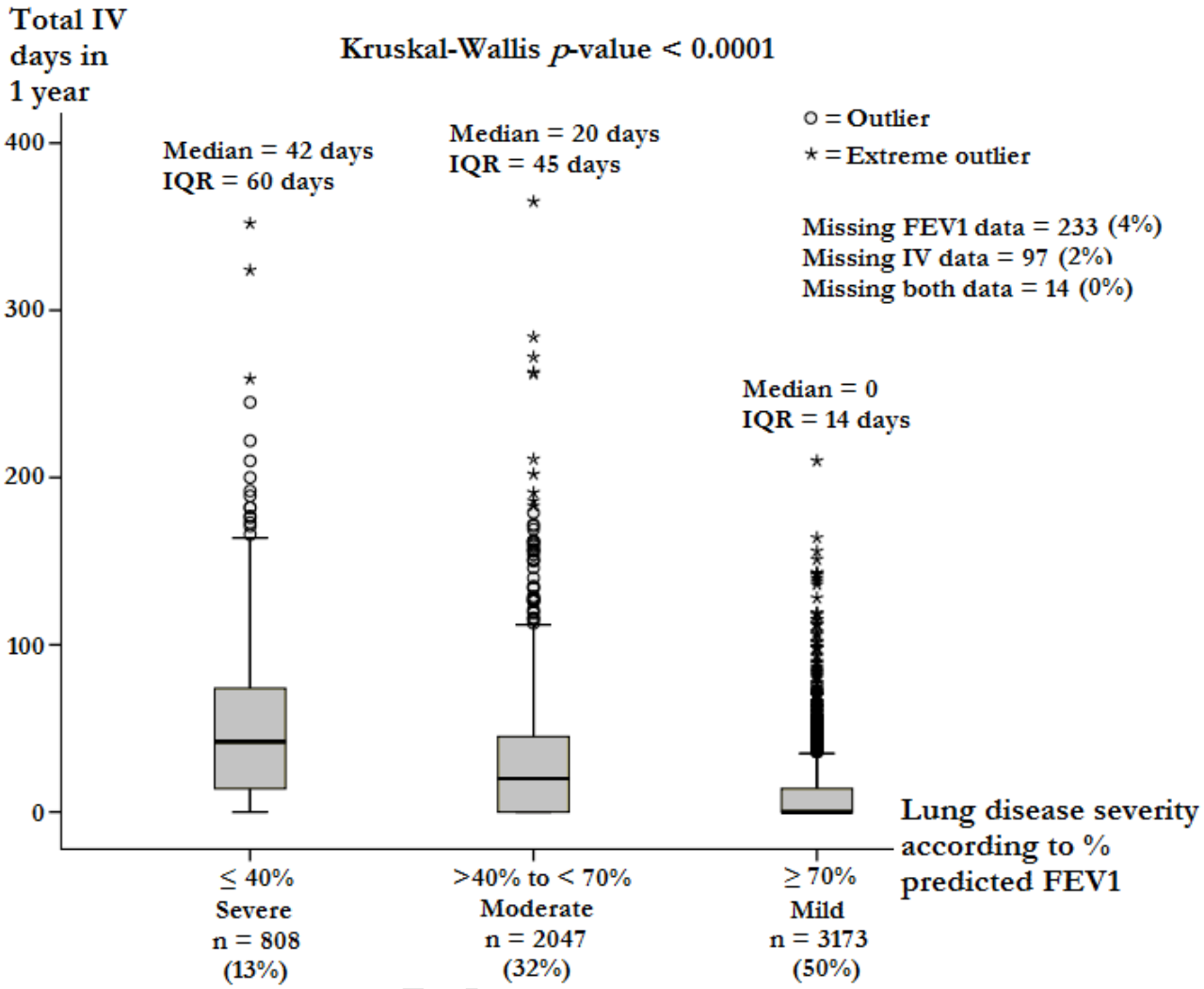

Fig. 2. Airway clearance techniques (*n*=6372). PWCF, people with cystic fibrosis; PEP, peak expiratory pressure.

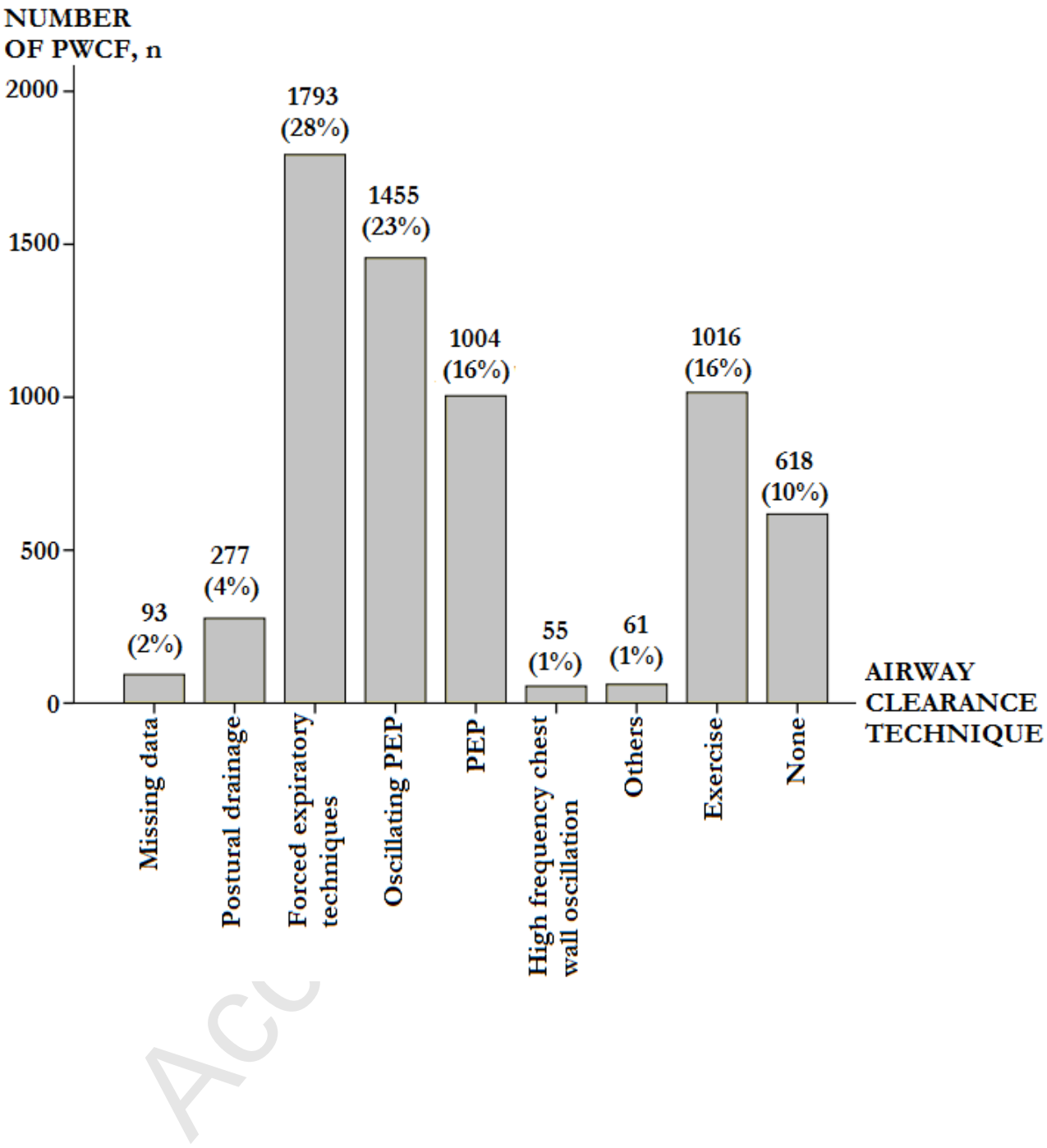

Table 1

Demographic details for people with cystic fibrosis (CF) included in this analysis (*n*=6372)

Age (years)	Median (IQR)	23 (15)
	Missing data (%)	0
Sex	Male, *n* (%)	3461 (54%)
	Female, *n* (%)	2911 (46%)
	Missing data, *n* (%)	0
FEV_1 (% predicted)	Median (IQR)	71.9 (37.1)
	Missing data (%)	247 (4%)
Total intravenous antibiotic-days	Median (IQR)	14 (37)
	Missing data (%)	111 (2%)
Use of DNase	Yes, *n* (%)	3387 (53%)
	No data[a], *n* (%)	2985 (47%)
Use of nebulised hypertonic sodium chloride	Yes, *n* (%)	1205 (19%)
	No data[a], *n* (%)	5167 (81%)

IQR, interquartile range; FEV_1, forced expiratory volume in 1 second.

[a]Data for use of DNase and nebulised hypertonic sodium chloride were collected by the UK CF Registry with a check-box with no option for 'not using'. Therefore, if the check-box was not selected (i.e. 'no data'), it could be a person with CF was not using that mucolytic/osmotic, but it could also represent missing data.

Table 2

Proportion of people with cystic fibrosis using different airway clearance techniques

Airway clearance techniques	Severe lung disease (% FEV_1 ≤40%) *n*=811	Moderate lung disease (% FEV_1 >40% to <70%) *n*=2052	Mild lung disease (% FEV_1 ≥70%) *n*=3181
Postural drainage	37 (5%)	95 (5%)	127 (4%)
Forced expiratory techniques	332 (41%)	630 (33%)	727 (23%)
Oscillating PEP	184 (23%)	485 (23%)	727 (23%)
PEP	84 (10%)	288 (14%)	592 (19%)
High-frequency chest wall oscillation	14 (2%)	18 (1%)	20 (1%)
Other	17 (2%)	19 (1%)	24 (1%)
Exercise	78 (10%)	311 (15%)	607 (19%)
None	65 (8%)	166 (8%)	357 (11%)

PEP, positive expiratory pressure.

Chi-square test *P*-value comparing airway clearance technique by severity of lung disease <0.0001.

None of the cells had an expected count <5. The minimum expected count was 7.

Missing data=328 (5%).

Table 3

Proportion of people with cystic fibrosis using nebulised mucolytic/osmotic to help with airway clearance

	Severe lung disease (% FEV_1 ≤40%) *n*=824	Moderate lung disease (% FEV_1 >40% to <70%) *n*=2074	Mild lung disease (% FEV_1 ≥70%) *n*=3227
DNase			
No data	244 (30%)	769 (37%)	1848 (57%)
Yes	580 (70%)	1305 (63%)	1379 (43%)
Nebulised hypertonic sodium chloride			
No data	607 (74%)	1586 (76%)	2776 (86%)
Yes	217 (26%)	488 (24%)	451 (14%)

FEV_1, forced expiratory volume in 1 second.

Chi-square test *p*-value comparing use of DNase by severity of lung disease <0.0001

Chi-square test *p*-value comparing use of nebulised hypertonic sodium chloride by severity of lung disease <0.0001

None of the cells had an expected count <5. For use of DNase, minimum expected count was 385. For use of nebulised hypertonic sodium chloride, minimum expected count was 156.

Missing data=247 (4%).

Table 4

Comparison of airway clearance technique use in the UK with other countries

	UK data 2011 (*n*=6372) ≥11 years	New Zealand data 2011 (*n*=138) ≥16 years	Canada data 2011 (*n*=3913) All ages	Canada data 2008 (*n*=2363) All ages	Ireland data (*n*=388) Adult data	US data (*n*=204)[a] Adult data	Argentina data (*n*=110)[a] Child data
Postural drainage	4%	2%	N/A	20%	0%	19%	54%
Forced expiratory techniques (including huff, ACBT and AD)	28%	14%	7%	5%	3% (ACBT) 46% (AD)	25%	33%
Oscillating PEP	23%	17%	6%	8%	17%	15%	11%[b]
PEP	16%	23%	34%	40%	10%		
High frequency chest wall oscillation	1%	2%	7%	3%	4%	37%	
Exercise	16%	46%					
Percussion			29%		0%		
Other	1%	0%	9%		3%		
None	10%	4%	8%				
Data source/collection method	UK CF registry database 2011	New Zealand registry report 2011 [19]	Canadian registry report 2011 [30]	McIlwaine *et al.* 2008 [18] using records from 24 CF clinics	CF registry of Ireland 2011 [20]	Sawicki *et al.* 2009 [17] using questionnaire as part of project on Adult Care in CF study	Ratto *et al.* 2012 [16] – Records from a single CF clinic

ACBT, active cycle of breathing techniques; AD, autogenic drainage; PEP, peak expiratory pressure; CF, cystic fibrosis.

[a]More than one airway clearance technique may be recorded if a person with cystic fibrosis used more than one technique.

[b]11% PEP and oscillating PEP.