

Positive expiratory pressure (PEP) therapy. What pressures do we achieve in young children with cystic fibrosis? A single-centre study

Niamh Kiernan ,¹ Barry Johnstone,^{1,2} Peter Anderson,¹ Ruth Stewart¹

To cite: Kiernan N, Johnstone B, Anderson P, *et al.* Positive expiratory pressure (PEP) therapy. What pressures do we achieve in young children with cystic fibrosis? A single-centre study. *BMJ Paediatrics Open* 2020;**4**:e000792. doi:10.1136/bmjpo-2020-000792

Received 22 July 2020
Accepted 3 December 2020

ABSTRACT

This study was a clinical review of infant positive expiratory pressure (PEP) therapy in young children with cystic fibrosis (CF). The aim of this study was to determine whether pressures of 10–20 cm H₂O PEP therapy (recommended by the CF trust) are being achieved with routine airway clearance therapy. This took place at the Royal Hospital for Children, Glasgow a specialist UK CF centre. Values were obtained from 21 young children. Pressures above 10 cm H₂O during tidal volume breathing were not achieved within our cohort. Further investigation is required to determine efficacy of lower pressures in PEP therapy with young children.

Airway clearance therapy (ACT) is a key aspect of the management in cystic fibrosis (CF).¹ CF lung disease is characterised by a cycle of infection, inflammation and airway obstruction which may progress to bronchiectasis.² ACT aims to clear mucus within the airways to slow progression of lung disease.³

Several forms of ACT are used for individuals with CF, however, no form has been shown to be superior. UK CF standards on ACT state that ‘all CF patients should be considered for positive expiratory pressure (PEP) therapy’. The guidance further recommends that for young children the appropriate resistance is one which ‘achieves a stable mid-expiratory pressure of 10–20 cm H₂O’.⁴ This recommendation is based on evidence from adult cohorts and minimal published data is available for young children. At the Royal Hospital for Children Glasgow, babies newly diagnosed with CF begin a daily ACT regimen involving PEP therapy via a fitted mask and assisted autogenic drainage (AAD). Therapy is performed on the parent’s knee with 2 min alternating cycles of PEP and AAD (four times).⁵ The initial PEP resistor diameter is set to 3.5 mm for term babies. This has been determined through clinical experience; to achieve an increase in resistance without a

significant increase in work of breathing. At routine reviews, resistor size is adjusted using clinical judgement.

The aim of this study was to measure pressures achieved by young children using PEP therapy in a CF cohort at a UK specialist paediatric CF centre.

A clinical review of pressures achieved during PEP therapy in young children attending a specialist CF centre was conducted. Children under 4 years of age receiving PEP mask therapy as part of their routine care were evaluated while parents performed their ACT on their knee. Data were collected by the physiotherapist using a digital manometer and measured over a cycle of five breaths with the peak pressures being recorded. Observations made in each child were recorded as; settled normal breathing, settled forced breathing, crying and settled but with hiccups.

Twenty-one young children with CF (female=10) were included in the study. Median age was 17 months median age is 17 months (IQR 6–22 months). Initial resistor sizes ranged from 2.5 to 3.5 mm. A mean pressure of 3.74 cm H₂O (range 0.3–16.18 cm H₂O) were recorded across the cohort. [Table 1](#) shows further details of data collected and [figure 1](#) shows a scatter plot of pressures achieved with increased age. A subset of six children had the test repeated using a 1.5 mm resistor. This was performed as part of their clinical management to identify whether using a higher resistance would achieve a higher expiratory pressure. Two of these children showed an increase of greater than 2 cm H₂O with the remaining four having negligible differences ([table 1](#)).

Young children achieved an average pressure of 3.74 cm H₂O with their routine PEP therapy. Values above 10 cm H₂O during tidal volume breathing were not achieved within



© Author(s) (or their employer(s)) 2020. Re-use permitted under CC BY-NC. No commercial re-use. See rights and permissions. Published by BMJ.

¹Respiratory Physiotherapy, Royal Hospital for Children, Glasgow, Scotland, UK

²Physiotherapy, NHS Greater Glasgow and Clyde, Glasgow, UK

Correspondence to

Miss Niamh Kiernan;
nkiernan89@outlook.com

Table 1 Summary of patient's age, PEP mask with resistor and PEP values achieved

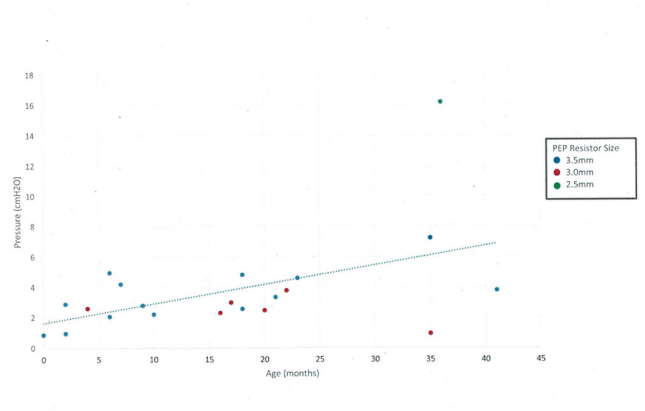
Subject	Resistor size (mm)	Breathing	Age (months)	Mask	Mean pressure (cm H ₂ O)	Pressure range (cm H ₂ O)
1	3.5	Settled	35	Pari size 3	7.22	5.6–9.6
2	3.5	Settled	2	Pari size 1	2.88	1.3–3.8
3	3.5	Settled	18	Pari size 2	2.56	2.2–2.9
4	3	Settled, hiccups	20	Pari pep 3	2.46	1.2–4.2
5	3.5	Settled	10	Pari size 2	2.2	2–2.6
6	3	Settled	41	Pari size 2	3.78	1.4–5.7
7	2.5	Settled (parents told force breath)	36	Pari size 2	16.18	7.1–23.2
8	3	Crying	4	Pari size 2	2.6	1.7–3.8
8	1.5	Crying	4	Pari size 2	2.5	0.7–6.4
9	3	Settled	35	Vital size toddler 3	9.4	0.3–1.3
9	1.5	Settled	35	Vital size toddler 3	1.7	1.2–2.7
10	3.5	Settled	6	Pari size 1	2.06	1.5–2.4
11	3	Settled	22	Vital size toddler 3	3.78	3.4–4.1
12	1.5	Settled	21	Pari size 2	3.28	1.4–5.0
12	3.5	Settled	21	Pari size 2	3.34	1.9–4.3
13	3	Settled	17	Pari size 2	2.98	2.2–3.2
14	3.5	Settled (forced)	18	Quadralite white	13.98	11.4–16.2
14	3.5	Settled (normal breathing)	18	Quadralite white	4.82	3.8–7.7
15	3.5	Settled	23	Pari size 2	4.6	3.5–5.9
16	3.5	Crying	7	Pari size 2	4.2	3.1–6.4
16	1.5	Crying	7	Pari size 2	6.26	2.5–12.2
17	3.5	Settled	2	Pari size1	0.96	0.3–2.0
18	3	Crying	16	Pari size 2	2.3	0.9–3.3
19	3.5	Crying	9	Pari size 2	2.8	2.4–3.4
20	3.5	Crying	6	Pari size 2	4.96	2.4–10.6
20	1.5	Crying	6	Pari size 2	7.28	2.0–13.3
21	3.5	Crying	0	Pari size 1	0.86	0.5–1.2
21	1.5	Crying	0	Pari size 1	0.84	0.0–2.2

PEP, positive expiratory pressure.

our cohort. Children are known to have higher airway resistance with higher chest wall compliance.² It could therefore be hypothesised that much lower PEP pressures may be effective. Furthermore, this study found that

increasing age or changing resistor size does not relate to achieving higher pressures. One older child (subject 7) was able to achieve higher pressures above 10 cm H₂O but only when prompted by a parent to 'blow harder'. Switching to a 'more active' PEP device when the child is able to (around 3 years of age) may be more effective in achieving higher consistent PEP pressures.⁶

Further work is required to establish the efficacy of these lower pressures in young children with CF.

**Figure 1** Scatterplot age versus pressure.

PATIENT AND PUBLIC INVOLVEMENT

Patients and/or the public were not involved in the design, or conduct, or reporting, or dissemination plans of this research.

Twitter Niamh Kiernan @niamh_kiernan and Barry Johnstone @barryjohnstone9

Contributors PA, RS and NK conceived of the presented idea. PA developed the theory and NK and PA performed the computations. BJ and NK verified the data



and formatted the graphs. PA encouraged NK to investigate previous work and standard. All authors discussed the results and contributed to the final manuscript.

Funding The authors have not declared a specific grant for this research from any funding agency in the public, commercial or not-for-profit sectors.

Competing interests None declared.

Patient consent for publication Not required.

Provenance and peer review Not commissioned; externally peer reviewed.

Open access This is an open access article distributed in accordance with the Creative Commons Attribution Non Commercial (CC BY-NC 4.0) license, which permits others to distribute, remix, adapt, build upon this work non-commercially, and license their derivative works on different terms, provided the original work is properly cited, appropriate credit is given, any changes made indicated, and the use is non-commercial. See: <http://creativecommons.org/licenses/by-nc/4.0/>.

ORCID iD

Niamh Kiernan <http://orcid.org/0000-0002-2887-2869>

REFERENCES

- 1 Warnock L, Gates A, Van der Shans C. Chest physiotherapy compared to no chest physiotherapy for cystic fibrosis. *Cochrane Database Syst Rev* 2015:CD001401.
- 2 McIlwaine M, Bradley J, Elborn JS, *et al*. Personalising airway clearance in chronic lung disease. *Eur Respir Rev* 2017;26. doi:10.1183/16000617.0086-2016. [Epub ahead of print: 21 Feb 2017].
- 3 Lannefors L, Button BM, McIlwaine M. Physiotherapy in infants and young children with cystic fibrosis: current practice and future developments. *J R Soc Med* 2004;97(Suppl 44):8–25.
- 4 Cystic Fibrosis Trust. *Standards of care and good clinical practice for the physiotherapy management of cystic fibrosis*, 2017.
- 5 Corten L, Jelsma J, Human A, *et al*. Assisted autogenic drainage in infants and young children hospitalized with uncomplicated pneumonia, a pilot study. *Physiother Res Int* 2018;23:e1690.
- 6 Carella E, Johnstone B, Brown S. 242 effectiveness of universal tubing at generating positive expiratory pressure in children: a pilot study. *J Cyst Fibros* 2011;10:S61.