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Lung volume recruitment and airway clearance for children at home in France

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Keywords:

Airway clearance; Cough; Lung volume recruitment; Pediatrics; Respiratory physiotherapy; Thoracic expansion.

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Abstract

Background:

Airway clearance (ACT) and lung volume recruitment (LVR) techniques are used to manage bronchial secretions, increase cough efficiency and lung/chest wall recruitment, to prevent and treat respiratory tract infections. The aim of the study was to review the prescription of ACT/LVR techniques for home use in children in France.

Methods:

All the centers of the national pediatric noninvasive ventilation (NIV) network were invited to fill in an anonymous questionnaire for every child aged ≤ 20 years who started a treatment with an ACT/LVR device between 2022 and 2023. The devices comprised mechanical in-exsufflation (MI-E), intermittent positive pressure breathing (IPPB), intrapulmonary percussive ventilation (IPV), and/or invasive mechanical ventilation (IMV)/NIV for ACT/LVR.

Results:

One hundred and thirty-nine patients were included by 13 centers. IPPB was started in 83 (60 %) patients, MI-E in 43 (31 %) and IPV in 30 (22 %). No patient used IMV/NIV for ACT/LVR. The devices were prescribed mainly by pediatric pulmonologists (103, 74 %). Mean age at initiation was 8.9 ± 5.6 (0.4-18.5) years old. The ACT/LVR devices were prescribed mainly in patients with neuromuscular disorders ($n = 66$, 47 %) and neurodisability ($n = 37$, 27 %). The main initiation criteria were cough assistance (81 %) and airway clearance (60 %) for MI-E, thoracic mobilization (63 %) and vital capacity (47 %) for IPPB, and airway clearance (73 %) and repeated respiratory exacerbations (57 %) for IPV. The parents were the main carers performing the treatment at home.

Conclusions:

IPPB was the most prescribed technique. Diseases and initiation criteria are heterogeneous, underlining the need for studies validating the indications and settings of these techniques.

1. Introduction

Patients with diseases that may impair the respiratory muscle function or alter the chest wall and lung mechanics, may present difficulties with airway clearance and/or cough efficacy [1–5]. This exposes to an increased risk of respiratory infection which may be the consequence of the inability to clear proximal or peripheral airway secretions because of muscle weakness, and/or repeated aspiration due to bulbar muscle dysfunction, and/or lung hypoventilation with collapsed lung areas [1, 6–8].

Airway clearance techniques (ACTs) are used to clear bronchial secretions, or assist or “replace” the cough function, and can be performed either by manual respiratory physiotherapy, or using specific devices, or both [3]. Proximal ACTs aim to augment cough efficiency by supporting the inspiratory phase, the expiratory phase or both [1]. Peripheral ACTs aim to increase secretion mobilization from the peripheral airways to the central airways, and improve ventilation. Some of these techniques have the advantages to be feasible in non-cooperative patients [1, 7, 9].

On the other hand, lung volume recruitment (LVR) techniques are used to increase alveolar recruitment and maintain lung and chest wall elasticity, in order to prevent respiratory tract infections, pulmonary atelectasis and ideally slow pulmonary function decline [1, 3, 8]. LVR can also be used as an inspiratory support to enhance cough efficacy [3, 8].

Several guidelines have been proposed to manage the respiratory airways in pediatrics, mainly for neuromuscular diseases (NMD) or pulmonary diseases (Pulmo) [3–5, 9–16]. However, recommendations are lacking regarding the indications and efficacy of home ACT/LVR in other disorders.

Only a few studies reported the use of some ACTs and LVR techniques in monocentric, multicentric or nationwide populations [2, 17, 18]. However, to our knowledge no study assessed the use of the different home ACT/LVR devices in a nationwide population. In France, the number of children that may require ACT/LVR and their disorders are not known. The aim of our study was to review the prescription of ACTs and LVR techniques, using specific devices at home, in the different centers of the French pediatric noninvasive ventilation (NIV) network, with the analysis of the initiation criteria, techniques, and settings of the devices in pediatric populations. We thought it was valuable to collect these data to better understand the national clinical practice, identify the populations using these devices, and potentially consider to propose French recommendations.

2. Material and methods

This study is a national prospective survey performed among the centers of the French national pediatric NIV network which gathers 28 pediatric university hospitals distributed among 24 cities [19]. All the centers were invited to fill in an anonymous questionnaire for every child aged ≤ 20 years who started a treatment with a device for ACTs and/or LVR within a 1-year period, between June 2022 and June 2023. Some young adults aged 18–20 years who were still followed in these centers since childhood, and who could not be followed by adult centers or were not yet transitioned, were also included. The survey was addressed to the physicians and respiratory therapists who follow patients with respiratory infections.

The mechanical ACT/LVR techniques consist of devices that deliver mechanical in-exsufflation (MI-E), intermittent positive pressure breathing (IPPB), intrapulmonary percussive ventilation (IPV), and/or devices for invasive mechanical ventilation (IMV)/NIV with settings for respiratory physiotherapy.

The questionnaire, which consists mainly on multiple choice questions and few items with open ended questions, comprised the following information: date of birth, gender, primary and secondary diagnosis, main therapies related to the disease, investigations performed before the treatment initiation, criteria that led to the decision for the treatment, type of device and interface used, prescriber, prescribed settings, daily and weekly recommended usage, use of additional equipment such as abdominal girdle and/or supplemental oxygen therapy, and additional treatments such as NIV or IMV and mouthpiece ventilation (MPV), and the carers involved in the treatment use. The data were collected from the clinical charts.

The investigations that could be performed before the ACT/LVR initiation included poly(somno)graphy (P(S)G), nocturnal gas exchange, pulmonary function tests (PFT), peak expiratory flow (PEF), cough peak flow (CPF), maximal static respiratory pressures (MRP), chest X-ray, or others.

The primary conditions leading to the prescription of ACT and/or LVR were grouped into 6 main diagnostic groups: NMD, neurodisability (Neurodis), scoliosis, Pulmo (i.e. lung diseases), thoracic diseases (Tho), spinal cord injuries (SCI), metabolic diseases (Metabo), and other disorders (Other). The questionnaires were centralized and analyzed by the NIV center of Necker hospital, in Paris. The study was approved by the institutional board (CERAPHP Centre) on June 2022, and all the patients aged >6 years and the parents gave their informed consent.

Descriptive statistics summarized the patient's characteristics and all the data related to the treatment.

Table 1
Patients' characteristics according to the type of ACT and LVR devices.

	All (n = 139)	MI-E (n = 43)	IPPB (n = 83)	IPV (n = 30)
Age at initiation (years)	9 ± 6 (0.4–19)	9 ± 6 (0.4–17)	9 ± 6 (0.4–19)	8 ± 6 (0.5–18)
M/F, n (%)	79 (57 %)/60 (43 %)	24 (56 %)/19 (44 %)	50 (60 %)/33 (40 %)	16 (53 %)/14 (47 %)
Primary disorder, n (%)				
NMD	66 (47 %)	22 (51 %)	47 (57 %)	5 (17 %)
Neurodis	37 (27 %)	17 (40 %)	8 (10 %)	19 (63 %)
Scoliosis	12 (9 %)	0	12 (14 %)	0
Pulmonary	7 (5 %)	0	3 (4 %)	4 (13 %)
Thoracic ^a	7 (5 %)	0	7 (8 %)	0
SCI	5 (4 %)	2 (5 %)	4 (5 %)	0
Metabo	4 (3 %)	2 (5 %)	1 (1 %)	2 (7 %)
Other disorders	1 (1 %)	0	1 (1 %)	0
Therapy for primary disease, n (%)				
None	113 (81 %)	30 (70 %)	68 (82 %)	25 (83 %)
Gene therapy	11 (8 %)	5 (12 %)	8 (10 %)	1 (3 %)
SMA				
Nusinersen	4 (3 %)	2 (5 %)	3 (4 %)	1 (3 %)
Risdiplam	3 (2 %)	2 (5 %)	1 (1 %)	1 (3 %)
Corticosteroids	2 (1 %)	1 (2 %)	1 (1 %)	0
Other ^b	6 (4 %)	3 (7 %)	2 (2 %)	2 (7 %)

Data are presented as mean ± standard deviation (range), or numbers (percentage of patients).

Abbreviations: ACT, airway clearance technique; LVR, lung volume recruitment; MI-E, mechanical in-exsufflation; IPPB, intermittent positive pressure breathing; IPV, intrapulmonary percussive ventilation; M, male; F, female; NMD, neuromuscular disorders; Neurodis, neurodisability; Pulmonary, pulmonary disorders; Thoracic, thoracic diseases; SCI, spinal cord injuries; Metabo, metabolic diseases; SMA, spinal muscular atrophy.

^a Poland syndrome.

^b Inhaled corticoids, bronchodilator, amifampridine, alglucosidase alfa, or rituximab.

3. Results

3.1. Patients and devices

One hundred and thirty-nine patients were included by 13 centers within the study period (Online Table 1). Some centers did not include patients, because the devices were not

indicated in their population of patients. Some patients were not included because of organizational issues. IPPB was started in 83 (60 %) patients, MI-E in 43 (31 %) patients and IPV in 30 (22 %) patients (Table 1). No patient had IMV/NIV with settings for ACT/LVR. Seventeen (12 %) patients were started on 2 techniques: 10 had a MI-E and IPV, and 7 had a MI-E and IPPB. Most of these 17 patients had spinal muscular atrophy type 1 or 2, or encephalopathy. The prescription of the devices was variable among centers (Table 2).

Table 2
Prescription of the ACT/LVR devices per center.

Center	Number of devices prescribed	ACT/LVR devices (n, %)		
		IPPB (n = 83)	MI-E (n = 43)	IPV (n = 30)
1	35	16 (46 %)	5 (14 %)	14 (40 %)
2	18	18 (100 %)	0	0
3	17	5 (29 %)	4 (24 %)	8 (47 %)
4	14	11 (79 %)	1 (7 %)	2 (14 %)
5	13	7 (54 %)	6 (46 %)	0
6	13	3 (23 %)	6 (46 %)	4 (31 %)
7	12	2 (17 %)	9 (75 %)	1 (8 %)
8	9	5 (56 %)	3 (33 %)	1 (11 %)
9	8	6 (75 %)	2 (25 %)	0
10	6	1 (17 %)	5 (83 %)	0
11	5	5 (100 %)	0	0
12	5	3 (60 %)	2 (40 %)	0
13	1	1 (100 %)	0	0

Data are presented as numbers and percentages of ACT/LVR devices prescribed in each participating center.

Abbreviations: ACT, airway clearance technique; LVR, lung volume recruitment; MI-E, mechanical in-exsufflation; IPPB, intermittent positive pressure breathing; IPV, intrapulmonary percussive ventilation.

The majority of patients (109, 78 %) had no prior ACT and/or LVR device, while 14 (10 %) patients had already an IPPB, 10 (7 %) patients had a MI-E, 4 (3 %) patients had NIV for physiotherapy and 2 (1 %) patients an IPV. The devices were prescribed mainly by a pediatric pulmonologist (103, 74 %), followed by a pediatric neurologist (24, 17 %), and a pediatric or adult rehabilitative physician (7, 5 %). Only one new device was prescribed per patient even in case of the initiation of 2 ACT/LVR techniques, as some devices combine 2 modalities (MI-E and IPPB or MI-E and IPV).

Table 1 presents the main characteristics of the patients according to the type of device. The ACT/LVR devices were mainly prescribed in patients with NMD (n = 66, 47 %), followed by Neurodis patients (n = 37, 27 %) and patients with scoliosis (n = 12, 9 %). Mean age at

initiation was 8.9 ± 5.6 (0.4–18.5) years old. The patients on IPV tended to be slightly younger than the others.

Twenty-nine patients were younger than 3 years old (16 were started on IPPB, 7 on MI-E and 9 on IPV), among which 6 were younger than 1 year old (2 were started on IPPB, 2 on MI-E and 3 on IPV). They had mainly NMD (spinal muscular atrophy (SMA) type 1 or 2, congenital myopathy) or thoracic diseases (Poland syndrome).

MI-E was prescribed preferentially in NMD and Neurodis patients, IPPB preferentially in NMD and scoliosis, and IPV in Neurodis, NMD and Pulmo patients. Most of the patients (113, 81 %) had no specific therapy for their primary disease, while 11 (8 %) patients received gene therapy for SMA.

Table 3
Investigations and initiation criteria for ACT and LVR treatments.

	MI-E (n = 43)	IPPB (n = 83)	IPV (n = 30)
Investigations, n (%)			
Lung function tests	13 (30 %)	53 (64 %)	0
Nocturnal gas exchange	18 (42 %)	20 (24 %)	9 (30 %)
Chest X-ray	17 (40 %)	21 (25 %)	13 (43 %)
PEF/CPF	8 (19 %)	13 (16 %)	0
Maximal static respiratory pressures	6 (14 %)	6 (7 %)	0
Poly(somno)graphy	3 (7 %)	6 (7 %)	1 (3 %)
None	14 (33 %)	10 (12 %)	11 (37 %)
Initiation criteria, n (%)			
Clin. Cough assistance	35 (81 %)	8 (10 %)	8 (27 %)
Clin. Airway clearance	26 (60 %)	20 (24 %)	22 (73 %)
Clin. Repeated respiratory exacerbations	20 (47 %)	11 (13 %)	17 (57 %)
Clin. Following ICU admission	7 (16 %)	1 (1 %)	4 (13 %)
Clin. Thoracic mobilization	1 (2 %)	52 (63 %)	1 (3 %)
Lung function test – VC	5 (12 %)	39 (47 %)	0
Clin. Atelectasis prevention	2 (5 %)	22 (27 %)	3 (10 %)
Clin. Atelectasis treatment	2 (5 %)	2 (2 %)	5 (17 %)
Lung function tests – Other ^a	0	16 (19 %)	0
PEF/CPF	3 (7 %)	4 (5 %)	0
Maximal static respiratory pressures	3 (7 %)	5 (6 %)	0
Prior to arthrodesis	1 (2 %)	14 (17 %)	3 (10 %)
Other	4 (1 %)	4 (5 %)	1 (3 %)

Data are presented as numbers (percentages of patients).

Abbreviations: ACT, airway clearance technique; LVR, lung volume recruitment; MI-E, mechanical in-exsufflation; IPPB, intermittent positive pressure breathing; IPV, intrapulmonary percussive ventilation; PEF, peak expiratory flow; CPF, cough peak flow; Clin., clinical indication; VC, vital capacity.

^a All tests performed were supine vital capacity.

3.2. Investigations and initiation criteria

A total of 227 investigations were performed prior to the treatment initiation. Fifty-nine (42 %) patients had one investigation, 26 (19 %) patients had 2 investigations and 17 (12 %) patients had 3 investigations. The main investigations performed were nocturnal gas exchange recording (42 %) and chest X-ray (40 %) in patients started on MI-E, PFT (64 %) for IPPB, and chest X-ray (43 %) for IPV (Table 3).

The main initiation criteria were cough assistance (81 %) and airway clearance (60 %) for MI-E, thoracic mobilization (63 %) and vital capacity (VC; 47 %) for IPPB, and airway clearance (73 %) and repeated respiratory exacerbations (57 %) for IPV (Table 3).

Mean VC in patients started on IPPB was 1.11 ± 0.62 (range 0.14–2.40) L and 60 ± 26 (range 13–118) % of predicted value.

3.3. Settings and other therapies

Table 4 summarizes the settings and conditions during the ACT/LVR device usage. The settings were adapted mainly by a respiratory physiotherapist alone (84 %), or by a physician alone (10 %).

For MI-E, mean expiratory pressure was higher than mean inspiratory pressure (Online Fig. 1), and mean expiratory time was higher than mean inspiratory time. Maximal pressures did not exceed 40 cmH₂O. Automatic mode was the most prescribed (84 %), and oscillations were rarely used (12 %).

For IPPB, maximal pressures were set at 40 cmH₂O (Online Fig. 1). Automatic mode was the most prescribed (67 %), and nebulization was rarely performed during the session (7 %).

For IPV, maximal pressures were set at 35 cmH₂O. Two or 3 different frequencies were set for the same session in 9 (30 %) patients, while the majority had only one frequency set. Nebulization was not prescribed, except for one patient.

Only one patient used oxygen during the treatment, because of a pneumopathy. The oronasal mask was the most common interface, while the mouthpiece was used only for IPPB in 41 % of the patients. Twenty-two percent of the patients used an abdominal girdle during the sessions, mainly during IPPB, and mainly in NMD patients. All the centers prescribed an abdominal girdle when appropriate.

Table 4
Settings and conditions of usage of ACT and LVR devices.

	All (n = 139)	MI-E (n = 43)	IPPB (n = 83)	IPV (n = 30)	
Settings^a	Pressures (cmH ₂ O)	PI 22 ± 7 (10, 40)	23 ± 6 (14, 40)	27 ± 7 (12, 35)	
		PE -26 ± 7 (-42, -10)			
	Airflow (lpm)		20 ± 6 (8, 30)		
	Time (sec)	Ti 1.7 ± 0.5 (1.0, 3.5)			
		Te 2.0 ± 0.6 (1.0, 4.0)			
		Tp 0.9 ± 0.5 (0.0, 2.0)			
Frequency (cpm)			566 ± 182 (300, 780)	328 ± 79 (200, 450)	
Other		Auto: n = 36 (84 %)	Rexp min: n = 68 (82 %) Auto: n = 56 (67 %)	Nebu: n = 1 (3 %)	
		Oscill: n = 5 (12 %)	Nebu: n = 6 (7 %)		
<i>Conditions during device use, n (%)</i>					
Oxygen therapy	1	0	1 ^b	0	
Abdominal girdle	31 (22 %)	8 (19 %)	23 (28 %)	1 (3 %)	
Oronasal mask	102 (73 %)	41 (95 %)	47 (57 %)	30 (100 %)	
Mouthpiece	34 (24 %)	0	34 (41 %)	0	

Data are presented as mean ± standard deviation (range), or numbers (percentage of patients).

Abbreviations: ACT, airway clearance technique; LVR, lung volume recruitment; MI-E, mechanical in-exsufflation; IPPB, intermittent positive pressure breathing; IPV, intrapulmonary percussive ventilation; PI, inspiratory pressure; PE, expiratory pressure; Ti, inspiratory time; Te, expiratory time; Tp, pause time; Auto, automatic mode; Oscill, oscillations; Rexp min, minimal expiratory resistance; Nebu, nebulization.

^a Ten missing data for MI-E and for IPV.

^b The patient had a pneumopathy.

Thirty-two (23 %) patients were already treated with long term nocturnal NIV or continuous positive airway pressure (CPAP), and 3 (2 %) were treated with IMV. Two other (1 %) patients had a tracheostomy without IMV. Six (4 %) patients had oxygen therapy and none used daytime MPV. The ACT/LVR devices were started concomitantly to nocturnal CPAP/NIV in 6 patients, or 17 ± 22 months after in 26 patients, and concomitantly to IMV in 1 patient or after IMV in 2 patients.

3.4. Usage and carers

Online Table 2 presents the recommended usage indications for the 3 devices. MI-E was recommended mainly on a needed basis daily (53 % of patients) and as often as needed during the week (58 %), while IPPB and IPV were recommended everyday (46 % and 37 %, respectively), once a day (92 % and 80 %, respectively).

The parents were the main carers who were performing the treatment at home (83 % of the patients), and they were the only carer for 57 % of the patients (Online Table 3). A respiratory physiotherapist performed the treatment either at the patient's follow-up care center (22 %) or at home (18 %). The respiratory physiotherapist was the only carer for only 16 % of the patients.

4. Discussion

Our prospective study analyzed the French practice of ACT and/or LVR techniques in pediatrics within tertiary university hospitals. During the study period, home ACT/LVR was initiated in 139 patients within 13 of the 28 centers of our national pediatric NIV network. The main patients' diseases were NMD, Neurodis and scoliosis. IPPB was the most prescribed technique, mainly for thoracic mobilization and LVR. The different devices could be started at a very young age, they were set mainly by a respiratory physiotherapist at the hospital, for a use principally at home by the parents.

To our knowledge, no study has reported the ACT and LVR indications and usage for children at home in France. Despite the limited number of disorders for which the French Health Insurance allows the reimbursement for the prescription of the devices (only for IPPB and MI-E), our findings highlight the fact that children with different disorders may necessitate an ACT/LVR device.

The number and type of devices prescribed by each center were different. However, our study showed that IPPB was the most prescribed technique (60 % of the patients), and could be used in very young children. IPPB was prescribed mostly for chest and lung expansion or to prevent atelectasis, and as an inspiratory assistance for coughing only in a few patients. IPPB was also prescribed in a few patients prior to spinal fusion surgery.

We did not report the short-term effects of IPPB on VC or the number of respiratory infections, as it was not the aim of the study. Only a few studies investigated the effects of a single session or regular use of IPPB for LVR, mainly in NMD [8,20–22]. But, according to the literature, the short-term or long-term benefits of LVR techniques, including IPPB, are still controversial [8]. The limited number of studies on IPPB may be explained by the fact that different techniques may be used for LVR [8]. To our knowledge, no study has evaluated the benefit of LVR prior to spinal surgery. Moreover, the optimal timing to start regular LVR is currently unknown and may differ according to the diseases [23]. In our study, the most common recommended usage for IPPB was once a day for a duration of 20 min. In the same way, there is a lack of guidelines regarding the optimal settings and recommended duration of use for LVR techniques. Regarding the role of IPPB/LVR for assisted inspiration prior to coughing, some studies reported that these techniques are effective to improve cough efficiency [1,9,24]. However, IPPB is not the first indication for assisted inspiration in children in France, probably because of the lack of convenience and the costs.

MI-E was prescribed in 31 % of the patients, comprising very young children. As in other studies [17,18], greater exsufflation pressures than insufflation pressures were usually set, with increased pressures with age (Online Fig. 1). Greater exsufflation than insufflation times were also usually set. Moreover, the devices were most often used with the automatic mode. However, in patients with neurodisability, exsufflation pressures were indeed higher than insufflation pressures ($\square 25 \pm 6$ vs 21 ± 7 cmH₂O), but mean exsufflation and insufflation times were similar (1.8 ± 0.4 vs 1.8 ± 0.6 s). Hov et al. [6] reported recently in children with neurodisability (including NMD), that symmetric settings resulted in better CPF, but that asymmetric or personalized settings led to better comfort scores. In the present study, we did not assess the comfort or CPF during MI-E, but the settings were adapted clinically and on the subjective comfort of the patient, as suggested by Hov et al. [6]. Some studies have given some guidance to adapt the settings for MI-E, but principally for NMD patients [25,26]. Guidelines are lacking for the chronic care of other populations, such as patients with neurodisability in whom MI-E may not be always recommended and pressure settings should be carefully adapted. Very few patients had a measurement of CPF, VC or MRP prior to MI-E initiation. However, as MI-E was used in many Neurodis patients, voluntary measurements may have been unfeasible. Moreover, in our study we did not report CPF measurements during the MI-E sessions. However, CPF was not found to correlate with the pressures required for an effective cough, questioning the real utility of CPF measurements to guide MI-E settings [17,27]. Other measurements such as electrical impedance tomography or pressure-flow curves could be helpful to better adapt MI-E settings, but further investigation is necessary to validate their value [26–28].

IPV was prescribed in 22 % of the patients, mainly to clear distal secretions in Neurodis patients. Interestingly, IPV was used less frequently than IPPB to prevent atelectasis [7]. To our knowledge, only one French study proposed some guidance for the settings of IPV [29],

but not for recent IPV devices. The lack of guidelines may be explained by the fact that this technique is not commonly used worldwide, and by the limited number of studies with small sample sizes [7].

Only 25 % of the patients were treated with home long term mechanical ventilation, and for more than half of the patients, home mechanical ventilation was not considered. None of the patients used home mechanical ventilation device for LVR or assisted inspiration for coughing, and only 3 % of the patients already had a “respiratory physiotherapy” program on their NIV devices. These findings highlight the fact that ACT/LVR techniques are most often prescribed independently of an IMV/NIV treatment. ACT/LVR should be always considered, when appropriate, in patients not candidates for mechanical ventilation or in whom it is contraindicated, in order to prevent respiratory infections [1, 3].

The parents were the only caregiver using ACT/LVR techniques in 57 % of the cases, which is representative of the real-life management of many children requiring ACT/LVR in France, either for practical reasons or because the parent can optimally manage the airway secretions or the LVR on his/her own. But this also probably highlights the lack of “home” respiratory physiotherapists trained to be able to use these devices in France, in order to lower the burden on the family carers.

Our study has several limitations. First, the number of patients is rather small, and all the patients were not included in the study due to logistical problems in some centers. Second, the diseases of the patients who were initiated on ACT/LVR may vary according to the centers due to the local recruitment of patients, with a center effect in some cases. Third, we did not analyze the center effect on the type of ACT/LVR device prescribed according to the initiation criteria and/or pathology, as the number of patients per center was too small. Therefore, our study did not allow to discuss the best indications and settings for the different devices. Fourth, this study analyzed only the initial prescription, without information on the real objective use of ACT/LVR, as many devices do not have adherence reports. Finally, the immediate effects of ACT/LVR or their long-term impact on the number of respiratory infections/exacerbations or hospitalizations were not reported, as it was not within the objectives of our study. We also did not gather the secretion burden and qualitative benefice of ACT/LVR devices.

5. Conclusions

This study shows that IPPB is the most common prescribed ACT/LVR technique in children at home in France, followed by MI-E and IPV. The age, diseases, and the initiation criteria are extremely heterogeneous, which makes it difficult to draw recommendations based on the French practice of these techniques. ACT/LVR devices may be successfully used in young children and children with neurodisability, even in case of poor cooperation. Simple measurements of interest using tools such as electrical impedance tomography, optoelectronic plethysmography, or pressure-flow curves, should be further investigated to guide the best settings of ACT/LVR techniques. Prospective studies are required to validate the indications, best timing and settings of the different ACT/ LVR devices in children according to their diseases, and to assess their efficacy.

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Ethics approval and consent to participate

Approved by the ethics committee CERAPHP Centre on June 2022 (No. 2022-01-09). Informed consent was obtained from all enrolled patients or their relatives.

CRedit authorship contribution statement

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Lucie Griffon: Writing – review & editing, Validation, Supervision, Investigation, Data curation.

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Declaration of competing interest

The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

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References

[1] M. Chatwin, M. Toussaint, M.R. Gonçalves, N. Sheers, U. Mellies, J. Gonzales- Bermejo, J. Sancho, B. Fauroux, T. Andersen, B. Hov, M. Nygren-Bonnier, M. Lacombe, K. Pernet, M. Kampelmacher, C. Devaux, K. Kinnett, D. Sheehan, F. Rao, M. Villanova, D. Berlowitz, B.M. Morrow, Airway clearance techniques in neuromuscular disorders: a state of the art review, *Respir. Med.* 136 (2018) 98–110.

[2] L. Rose, D. McKim, D. Leasa, M. Nonoyama, A. Tandon, M. Kaminska, C. O’Connell, A. Loewen, B. Connolly, P. Murphy, N. Hart, J. Road, Monitoring cough effectiveness and use of airway clearance strategies: a Canadian and UK survey, *Respir. Care* 63 (2018) 1506–1513.

[3] M. Toussaint, M. Chatwin, J. Gonzales, D.J. Berlowitz, ENMC respiratory therapy consortium, 228th ENMC International Workshop: airway clearance techniques in neuromuscular disorders Naarden, The Netherlands, 3-5 March, 2017, *Neuromuscul. Disord.* (2018) 3.

- [4] B. Herrero-Cortina, A.L. Lee, A. Oliveira, B. O'Neill, C. J'acome, S. Dal Corso, W. Poncin, G. Munoz, D. Inal-Ince, V. Alcaraz-Serrano, G. Reychler, A. Bellofiore, A. Posthumus, Patient representative, T. Tonia, J.D. Chalmers, A. Spinou, European Respiratory Society statement on airway clearance techniques in adults with bronchiectasis, *Eur. Respir. J.* 62 (2023) 2202053, 2202010.2201183/ 13993003.13902053-13992022.
- [5] P.A. Flume, K.A. Robinson, B.P. O'Sullivan, J.D. Finder, R.L. Vender, D.B. Willey-Courand, T.B. White, B.C. Marshall, Clinical practice guidelines for pulmonary therapies committee, cystic fibrosis pulmonary guidelines: airway clearance therapies, *Respir. Care* 54 (2009) 522–537.
- [6] B. Hov, T. Andersen, M. Toussaint, I.B. Mikalsen, M. Vollsæter, C. Brunborg, M. Hovde, V. Hovland, Mechanically assisted cough strategies: user perspectives and cough flows in children with neurodisability, *ERJ Open Res* 10 (2024) 274–2023, 00210.01183/23120541.23100274-23122023.
- [7] E. Lauwers, K. Ides, K. Van Hoorenbeeck, S. Verhulst, The effect of intrapulmonary percussive ventilation in pediatric patients: a systematic review, *Pediatr. Pulmonol.* 53 (2018) 1463–1474.
- [8] N.L. Sheers, R. O'Sullivan, M.E. Howard, D.J. Berlowitz, The role of lung volume recruitment therapy in neuromuscular disease: a narrative review, *Front Rehabil Sci.* 4 (2023) 1164628, 1164610.1163389/fresc.1162023.1164628.
- [9] J. Hull, R. Aniapravan, E. Chan, M. Chatwin, J. Forton, J. Gallagher, N. Gibson, J. Gordon, I. Hughes, R. McCulloch, R.R. Russell, A. Simonds, British Thoracic Society guideline for respiratory management of children with neuromuscular weakness, *Thorax* 67 (Suppl 1) (2012) i1–i40.
- [10] H.M. Sorenson, D.C. Shelledy, AARC clinical practice guideline. Intermittent positive pressure breathing–2003 revision & update, *Respir. Care* 48 (2003) 540–546.
- [11] M. Boentert, H. Prigent, K. Vardi, H.N. Jones, U. Mellies, A.K. Simonds, S. Wenninger, E. Barrot Cortés, M. Confalonieri, Practical recommendations for diagnosis and management of respiratory muscle weakness in late-onset pompe disease, *Int. J. Mol. Sci.* 17 (2016) 1735, 1710.3390/ijms17101735.
- [12] R.S. Finkel, E. Mercuri, O.H. Meyer, A.K. Simonds, M.K. Schroth, R.J. Graham, J. Kirschner, S.T. Iannaccone, T.O. Crawford, S. Woods, F. Muntoni, B. Wirth, J. Montes, M. Main, E.S. Mazzone, M. Vitale, B. Snyder, S. Quijano-Roy, E. Bertini, R.H. Davis, Y. Qian, T. Sejersen, SMA Care group, Diagnosis and management of spinal muscular atrophy: Part 2: pulmonary and acute care; medications, supplements and immunizations; other organ systems; and ethics, *Neuromuscul. Disord.* 28 (2018) 197–207.

- [13] J.D. Finder, D. Birnkrant, J. Carl, H.J. Farber, D. Gozal, S.T. Iannaccone, T. Kovesi, R.M. Kravitz, H. Panitch, C. Schramm, M. Schroth, G. Sharma, L. Sievers, J. M. Silvestri, L. Sterni, American Thoracic Society, Respiratory care of the patient with Duchenne muscular dystrophy: ATS consensus statement, *Am. J. Respir. Crit. Care Med.* 170 (2004) 456–465.
- [14] D.W. Sheehan, D.J. Birnkrant, J.O. Benditt, M. Eagle, J.D. Finder, J. Kissel, R. M. Kravitz, H. Sawnani, R. Shell, M.D. Sussman, L.F. Wolfe, Respiratory management of the patient with Duchenne muscular dystrophy, *Pediatrics* 142 (Suppl 2) (2018) S62–S71.
- [15] D.J. Birnkrant, K. Bushby, C.M. Bann, B.A. Alman, S.D. Apkon, A. Blackwell, L. E. Case, L. Cripe, S. Hadjiyannakis, A.K. Olson, D.W. Sheehan, J. Bolen, D. R. Weber, L.M. Ward, DMD Care Considerations Working Group, Diagnosis and management of Duchenne muscular dystrophy, part 2: respiratory, cardiac, bone health, and orthopaedic management, *Lancet Neurol.* 17 (2018) 347–361.
- [16] B.M. Button, C. Wilson, R. Dentice, N.S. Cox, A. Middleton, E. Tannenbaum, J. Bishop, R. Cobb, K. Burton, M. Wood, F. Moran, R. Black, S. Bowen, R. Day, J. Depiazzi, K. Doiron, M. Doumit, T. Dwyer, A. Elliot, L. Fuller, K. Hall, M. Hutchins, M. Kerr, A.L. Lee, C. Mans, L. O’Connor, R. Steward, A. Potter, T. Rasekaba, R. Scoones, B. Tarrant, N. Ward, S. West, D. White, L. Wilson, J. Wood, A.E. Holland, Physiotherapy for cystic fibrosis in Australia and New Zealand: a clinical practice guideline, *Respirology* 21 (2016) 656–667.
- [17] M. Chatwin, A.K. Simonds, Long-term mechanical insufflation-exsufflation cough assistance in neuromuscular disease: patterns of use and lessons for application, *Respir. Care* 65 (2020) 135–143.
- [18] B. Hov, T. Andersen, V. Hovland, M. Toussaint, The clinical use of mechanical insufflation-exsufflation in children with neuromuscular disorders in Europe, *Paediatr. Respir. Rev.* 27 (2018) 69–73.
- [19] B. Fauroux, S. Khirani, A. Amaddeo, B. Massenavette, P. Bierme, J. Taytard, N. Stremmer, M. Baravalle-Einaudi, J. Mazenq, I. Ioan, C. Schweitzer, M. Lampin, A. Binoche, C. Mordacq, J. Bergounioux, B. Mbieleu, R. Rubinsztajn, E. Sigur, G. Labouret, A. Genevois, A. Becourt, E. Hullo, I. Pin, S. Debelleix, F. Galod’*e*, S. Bui, J. Moreau, M. Renoux, S. Matecki, M. Lavadera, R. Heyman, M. Pomedio, L. Le Clainche, P. Bokov, A. Masson, P. Hangard, C. Menetrey, M. Jokic, E. Gachelin, C. Perisson, A. Pervillé, A. Fina, L. Giovannini-Chami, E. Fleurence, A. Barzic, A. Breining, M. Ollivier, G. Labbé, L. Coutier, G. Aubertin, Paediatric long term continuous positive airway pressure and noninvasive ventilation in France: a cross-sectional study, *Respir. Med.* 181 (2021) 106388, 106310.101016/j.rmed.102021.106388.
- [20] C. Guérin, B. Vincent, T. Petitjean, P. Lecam, C. Luizet, M. Rabilloud, J.C. Richard, The short-term effects of intermittent positive pressure breathing treatments on ventilation in patients with neuromuscular disease, *Respir. Care* 55 (2010) 866–872.

- [21] I. Laffont, D. Bensmail, S. Lortat-Jacob, L. Falaize, C. Hutin, E. Le Bomin, M. Ruquet, P. Denys, F. Lofaso, Intermittent positive-pressure breathing effects in patients with high spinal cord injury, *Arch. Phys. Med. Rehabil.* 89 (2008) 1575–1579.
- [22] A. De Troyer, P. Deisser, The effects of intermittent positive pressure breathing on patients with respiratory muscle weakness, *Am. Rev. Respir. Dis.* 124 (1981) 132–137.
- [23] N.L. Sheers, M.E. Howard, P.D. Rochford, L. Rautela, C. Chao, D.A. McKim, D. J. Berlowitz, A randomized controlled clinical trial of lung volume recruitment in adults with neuromuscular disease, *Ann Am Thorac Soc* 20 (2023) 1445–1455.
- [24] U. Mellies, C. Goebel, Optimum insufflation capacity and peak cough flow in neuromuscular disorders, *Ann Am Thorac Soc* 11 (2014) 1560–1568.
- [25] M. Chatwin, R.H. Wakeman, Mechanical insufflation-exsufflation: considerations for improving clinical practice, *J. Clin. Med.* 12 (2023) 2626, 2610.3390/jcm12072626.
- [26] C. Lalmolda, H. Prados, G. Mateu, M. Noray, X. Pomares, M. Luján, Titration of mechanical insufflation-exsufflation optimal pressure combinations in neuromuscular diseases by flow/pressure waveform analysis, *Arch. Bronconeumol.* 55 (2019) 246–251.
- [27] N.M. Shah, C. Apps, G. Kaltsakas, S. Madden-Scott, E.S. Suh, R.F. D’Cruz, G. Arbane, M. Patout, E. Lhuillier, N. Hart, P. Murphy, The effect of pressure changes during mechanical insufflation-exsufflation on respiratory and airway physiology, *Chest* S0012–3692 (23) (2023) 5572, 05571. doi: 05510.01016/j.chest.02023.05510.05015.
- [28] M. Lacombe, A. Boré, L.D. Amo Castrillo, G. Boussaïd, L. Falaize, E. Vlachos, D. Orlikowski, H. Prigent, F. Lofaso, Peak cough flow fails to detect upper airway collapse during negative pressure titration for cough-assist, *Arch. Phys. Med. Rehabil.* 100 (2019) 2346–2353.
- [29] G. Riffard, M. Toussaint, [Intrapulmonary percussion ventilation: operation and settings], *Rev. Mal. Respir.* 29 (2012) 347–354.