Table 3. Respiratory physiotherapy management at the end of follow-up

	med [Q1;Q3]	**(min;max)**
Mechanical Insufflator/Exsufflator (MI:E)		
Number of patients (n; %)	11 (61)	
Age at onset	14 [11;21]	(10;30)
Intra-Pulmonary Percussive Ventilation (IPPV)		
Number of patients (n; %)	3 (18)	
Age at onset	6 [5.5;8]	(5;10)
Intermittent positive pressure breathing (IPPB)		
Number of patients (n; %)	8 (47)	
Age at onset	25 [13.3;33.5]	(9;41)
Abdominal belt		
Number of patients (n; %)	10 (67)	
Age at onset	12 [9;19.5]	(5;30)
Oral Suctionning		
Number of patients (n; %)	13 (76)	
Age at onset	10 [5;16]	(2.5;35)

Results are expressed as median, interquartile range: med [Q1;Q3]; minimum and maximum (min;max). MI:E and IPPV are both airway secretion management devices. MI:E simulates a cough thanks to an insufflation phase (inflating the lungs) immediately followed by a change to negative pressure allowing a rapid exhalation (exsufflation phase). IPPV allows the mucus to move up the airway by producing small rapid bursts of air (percussions). IPPV use is followed by oral suctioning to remove the secretions. IPPB is used to provide controlled positive pressure via mouthpiece or face mask to promote ventilation, and thoracic and lung expansion. The abdominal belt is used to maintain the abdomen and preventing its distension during the use of Non-Invasive Ventilation and/or respiratory physiotherapy devices, to allow a maximum thoracic and lung expansion.