

## Cerebral palsy in adults

**[C2] Identifying and managing respiratory disorders associated with cerebral palsy: assisted ventilation**

*NICE guideline tbc*

*Evidence reviews*

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*These evidence reviews were developed by National Guideline Alliance hosted by the Royal College of Obstetricians and Gynaecologists*



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# 1 Assisted ventilation techniques for adults 2 with cerebral palsy

## 3 Review question

4 C2 Does assisted ventilation improve quality of life for adults with cerebral palsy who have a  
5 chronic respiratory disorder (including respiratory failure)?

## 6 Introduction

7 Conservative treatment for chronic respiratory disorders aims to ensure adequate supply of  
8 oxygenation. This is commonly achieved through treatment with bronchodilators,  
9 corticosteroids, and controlled oxygen. People who do not respond to such interventions  
10 receive ventilation. If ventilation involves any instrument that is inserted in the trachea  
11 through the mouth, such as an endotracheal tube, it is referred to as invasive assisted  
12 ventilation. This may be associated with adverse events such as tissue damage and  
13 infections. In non-invasive ventilation the patient receives air or a mixture of air and oxygen  
14 from a flow generator through a full facial or nasal mask which is a less invasive method of  
15 supplying sufficient oxygen to the body with potentially fewer adverse events. This evidence  
16 review sets out to investigate the effectiveness of assisted ventilation for adults with cerebral  
17 palsy who have a chronic respiratory disorder.

## 18 PICO Table

19 Please see Table 1 for a summary of the Population, Intervention, Comparison and Outcome  
20 (PICO) characteristics of this review.

21 **Table 1: Summary of the protocol (PICO table)**

<b>Population</b>	Adults aged 25 and over with cerebral palsy who have a chronic respiratory disorder (including respiratory failure)
<b>Intervention</b>	Assisted ventilation techniques: <ul style="list-style-type: none"><li>• Non-invasive ventilation (NIV)</li><li>• Ventilation via tracheostomy</li></ul>
<b>Comparison</b>	<ul style="list-style-type: none"><li>• No assisted ventilation</li><li>• NIV versus standard care</li><li>• NIV versus ventilation via tracheostomy</li></ul>
<b>Outcome</b>	<b>Critical</b> <ul style="list-style-type: none"><li>• Hospital admissions</li><li>• Overall survival</li><li>• Quality of life (carer or self-reported)</li></ul> <b>Important</b> <ul style="list-style-type: none"><li>• Treatment complications</li><li>• Daytime sleepiness and fatigue</li></ul>

22 *NIV: Non-invasive ventilation*

23 For full details see review protocol in appendix A.

## 24 Methods and process

25 This evidence review was developed using the methods and process described in  
26 [Developing NICE guidelines: the manual 2014](#). Methods specific to this review question are

1 described in the review protocol in appendix A and for a full description of the methods see  
2 supplementary document C.

3 Declaration of interests were recorded according to NICE's 2014 conflicts of interest policy  
4 from May 2016 until April 2018. From April 2018 onwards they were recorded according to  
5 NICE's 2018 [conflicts of interest policy](#). Those interests declared until April 2018 were  
6 reclassified according to NICE's 2018 conflicts of interest policy (see Interests Register).

## 7 **Clinical evidence**

### 8 **Included studies**

9 A systematic review of the clinical literature was conducted but no relevant studies were  
10 identified which were applicable to this review question.

11 See the literature search strategy in appendix B and study selection flow chart in appendix C.

### 12 **Excluded studies**

13 Studies excluded from this systematic review, with reasons for their exclusion, are provided  
14 in appendix K.

### 15 **Summary of clinical studies included in the evidence review**

16 No clinical studies were identified for this review.

### 17 **Quality assessment of clinical studies included in the evidence review**

18 No clinical studies were identified for this review.

## 19 **Economic evidence**

### 20 **Included studies**

21 A systematic review of the economic literature was conducted but no studies were identified  
22 which were applicable to this review question.

### 23 **Excluded studies**

24 No studies were identified which were applicable to this review question.

### 25 **Summary of studies included in the economic evidence review**

26 No economic evaluations were included in this review.

## 27 **Economic model**

28 Whilst this review topic was prioritised for bespoke economic modelling, the failure to identify  
29 any clinical evidence meant it would not have been feasible to model clinical and quality of  
30 life outcomes in a way which would have assisted in the making of recommendations. A  
31 discussion of costs have been presented in the resource impact section.

32

33

34

## 1 Resource impact

2 **Table 2: Unit cost for different types of assistive ventilation**

Resource	Unit costs†	Source
CPAP Device	£780 (£129-£7,898)	NHS Supply Catalogue 2017
Bi-level CPAP Device	£2,675 (£451-£7168)	NHS Supply Catalogue 2017
Tracheostomy (initial operation)	£2,972 (£175-£4,823)	Department of Health NHS Reference Costs 2016/2017

3 CPAP: Continuous positive airway pressure; NHS: National Health Service

4 † Median cost identified with the range of identified costs in parenthesis

### 5 **Continuous positive airway pressure (CPAP) and Bi-Level CPAP Devices**

6 CPAP and bi-level CPAP devices are full facial or nasal masks which ensure the adequate  
7 supply of oxygen to an individual. The main difference between CPAP and bi-level CPAP  
8 devices is that a bi-level CPAP device allows for 2 pressure settings for inhalation and  
9 exhalation making breathing easier and increasing the volume of air entering and exiting the  
10 lungs.

11 Costs for CPAP devices from NHS Supply Catalogue 2017 ranged from £129 to £7,898 with  
12 a median cost of £780. One previous UK economic evaluation was identified, NICE TA139  
13 (2008) which looked at [CPAP for the treatment of obstructive sleep apnoea/hypopnoea  
14 syndrome](#). This report estimated a cost of the CPAP device of £280. This cost does not  
15 report additional costs such as initial fitting of the mask, replacement parts and batteries for  
16 devices, additional equipment such as stands or backpacks and other sundries. Using an  
17 annual equivalent cost based on NICE methodology the report estimated the annual costs as  
18 £164.64 in the first year and £218.24 per person in subsequent years equivalent to £193.66  
19 and £256.71 in 2017 prices inflated using the hospital & community health services (HCHS)  
20 index of the Personal and Social Services Research Unit (PSSRU 2017). These estimates of  
21 costs for CPAP devices will likely represent a lower end for the cost of CPAP devices for  
22 adults with cerebral palsy. CPAP devices for cerebral palsy are likely to be used more  
23 frequently and there may be a need for the devices to be transported. This will increase  
24 replacement and maintenance costs and require additional equipment. Adults with cerebral  
25 palsy may also be likely to use more expensive CPAP devices with additional features such  
26 as breathing monitoring and alarms.

27 Costs for bi-level CPAP devices ranged from £451 to £7,168 in the NHS Supply Catalogue  
28 with a median cost of £2,675. This higher median cost compared to CPAP represents the  
29 increased sophistication of the bi-level machines. As above these costs did not include any  
30 additional costs although there was no reason to believe that these would differ to that of a  
31 CPAP device. One previous UK economic evaluation was identified, NICE NG42 (2016),  
32 which looked at non-invasive ventilation for people with [motor neurone disease](#). The report  
33 estimated a cost per machine of £4000 which, with the addition of other expenses, almost  
34 identical to NICE TA139 described above, and using identical methodology estimated an  
35 annual cost of £3,149.39 equivalent to £3440.77 in 2017 prices. It was considered that this  
36 would be in line with the cost for adults with cerebral palsy given that both patient groups  
37 would be using the devices for extended periods of the day, the need for additional  
38 equipment for transportation and relative sophistication of the devices.

### 39 **Ventilation via tracheostomy**

40 A tracheostomy is an operation where an incision is made in the trachea opening a direct  
41 airway. A tracheostomy costs between £175 and £4,823 with a median cost of £2,972  
42 depending on whether it is elective or non-elective or performed on an outpatient, day case  
43 or inpatient basis based on Department of Health Reference Costs 2016/2017. No previous

1 UK costings or economic evaluations were identified which considered ventilation via  
2 tracheostomy. Although the real cost drivers of ventilation by tracheostomy is the need for 24  
3 hour access to nursing, medical and technical support. The costs of this can be in the  
4 hundreds of thousands of pounds over the life course of the patient. Only a very small  
5 number of patients would opt for invasive ventilation over less costly non-invasive ventilation,  
6 not only because it is more invasive but also because the majority of patients receiving  
7 ventilation in this manner would be unable to receive ventilation by any other means.  
8 Therefore invasive and non-invasive ventilation should not be viewed as competing  
9 interventions. Patients in this group are also more likely to need treatment for infection (given  
10 the open trachea). They are also more likely to need emergency admission although this  
11 would be accounted for by this group having greater need for ventilation support rather than  
12 because of the intervention given. Both of these would lead to additional costs although they  
13 will be small relative to the other costs discussed.

#### 14 **Evidence statements**

#### 15 **Recommendations**

16 C2.1 If a person with cerebral palsy has symptoms of respiratory failure, or is at high risk of  
17 developing respiratory failure, discuss their management plan with them (and their family or  
18 carers if agreed), including:

- 19 • assessing the effectiveness and tolerability of treatment
- 20 • treatment goals and escalation plan of treatment
- 21 • managing complications
- 22 • options for managing progressive respiratory failure.

23 C2.2 Consider home-based non-invasive ventilation for adults with cerebral palsy and  
24 respiratory failure.

25 C2.3 If a person is having home-based non-invasive ventilation, carry out a review with a  
26 multidisciplinary team every 3 to 6 months. Assess and discuss with the person (and their  
27 family and carers, if agreed) the effectiveness, tolerability and whether agreed goals are  
28 being met.

29 C2.4 Discuss with the person having non-invasive ventilation (and their family or carers, if  
30 agreed) their preferences for future treatment. Agree a plan for what should happen if an  
31 intercurrent infection (occurring during the progress of respiratory failure) causes an acute  
32 deterioration in respiratory function and record this in the person's advance care plan.

33 C2.5 If the person's agreed treatment goals are not met by non-invasive ventilation, consider  
34 alternative treatment options such as tracheostomy or supportive care, taking into account  
35 the person's preferences for future treatment (see recommendation C2.4).

36 C2.6 If an adult with cerebral palsy has sleep apnoea follow the advice in the NICE  
37 technology appraisal guidance on [continuous positive airway pressure for the treatment of](#)  
38 [obstructive sleep apnoea/hypopnoea syndrome](#).

#### 39 **Rationale and impact**

#### 40 **Why the committee made the recommendations**

41 The management of respiratory failure varies according to individual circumstances and  
42 preferences. The committee noted that having discussions about the effectiveness and  
43 tolerability of treatments (for example, non-invasive ventilation can be uncomfortable), as  
44 well as planning for future treatment and what to do if their condition worsens, helps to  
45 identify the most appropriate treatment pathway.

1 Although no evidence was identified on assisted ventilation for adults with cerebral palsy, the  
2 committee noted that there is evidence supporting non-invasive ventilation in people with  
3 progressive neuromuscular conditions. The committee discussed that the course and  
4 symptoms of respiratory failure may be similar across the different conditions. They agreed  
5 that non-invasive ventilation could be beneficial, based on evidence extrapolated from these  
6 populations and the committee's experience. The committee also agreed that it is important  
7 to review management every 3–6 months, which is consistent with standard practice.

8 The committee discussed that people's goals and preferences for management after acute  
9 deterioration in respiratory function will vary. Based on their experience and expertise, the  
10 committee highlighted the importance of agreeing a management plan with the person (and  
11 their family or carers, if agreed) for future care in this situation. This should be documented in  
12 the person's advance care plan. A full understanding of the options available and the  
13 person's values, preferences and goals will lead to better shared decision-making and more  
14 informed choices about care.

15 Based on their experience and expertise, the committee discussed that when treatment  
16 goals are not met by non-invasive techniques, alternative options like tracheostomy or  
17 supportive care could be considered. There was no evidence available in adults with cerebral  
18 palsy, but the committee agreed that tracheostomy can be effective for some patients in  
19 maintaining quality of life.

20 The committee recognised that sleep apnoea is common in adults with cerebral palsy. It can  
21 affect sleep quality and therefore quality of life. They agreed that treatment would be the  
22 same for adults with cerebral palsy as in the general population and cross-referred to the  
23 NICE technology appraisal guidance on [continuous positive airway pressure for the  
24 treatment of obstructive sleep apnoea/hypopnoea syndrome](#).

## 25 **Impact of the recommendations on practice**

26 The recommendations in this section reinforce the current best practice and will help to  
27 standardise practice.

## 28 **The committee's discussion of the evidence**

### 29 **Interpreting the evidence**

#### 30 ***The outcomes that matter most***

31 The critical outcomes were hospital admissions, overall survival and quality of life, as  
32 assisted ventilation techniques are primarily aimed at improving these outcomes. To allow a  
33 balance of the benefits and harms of assisted ventilation techniques, treatment complications  
34 (for instance mask related difficulties) and daytime sleepiness and fatigue (when assistive  
35 ventilation led to sleep disturbance) were included as important outcomes.

#### 36 ***The quality of the evidence***

37 No evidence was identified for this review.

#### 38 ***Benefits and harms***

39 The committee discussed that people with cerebral palsy may have their own perception  
40 about quality of life and what may be considered as good quality of life by one individual,  
41 may not be considered acceptable by another. Based on their clinical experience, the  
42 committee discussed that some people may want to be put on assisted ventilation following  
43 acute deterioration in respiratory function, while others would not consider that as  
44 acceptable. The goals of management following acute deterioration in respiratory function  
45 may vary from individual to individual. The committee noted that this needs to be considered

1 while managing respiratory conditions. Hence the committee made the recommendation  
2 regarding agreeing a plan with person (and family or carers) in the event of infection causing  
3 acute deterioration in respiratory function, based on their clinical experience and expertise.  
4 The committee recognised that shared decision making will help the people achieve an  
5 acceptable level of quality of life. The committee agreed it was important to discuss a  
6 management plan with the adult with cerebral palsy who may be at risk to go on to develop  
7 respiratory complications. Such a plan would take into account the preferences of the person  
8 with cerebral palsy to balance the likely effectiveness of future treatment with its tolerability  
9 and impact on independence and family life (for example the impact that home assisted  
10 ventilation may have on daily routines).

11 There was no evidence about home based non-invasive ventilation for adults with cerebral  
12 palsy. Based on their knowledge the committee discussed that there is good evidence  
13 available regarding the effectiveness of non-invasive ventilation techniques for respiratory  
14 failure in progressive neuromuscular conditions (for instance the committee were aware of  
15 the recommendations in the NICE guideline on [motor neurone disease](#)). Considering that the  
16 course and symptoms of respiratory failure may be similar in people with cerebral palsy, the  
17 committee agreed that non-invasive ventilation could also benefit people with cerebral palsy.  
18 Due to the lack of direct evidence the committee decided to make a weak recommendation  
19 for this. The committee agreed that the technology and equipment for non-invasive  
20 ventilation would need to be checked and its effectiveness monitored. Based on their  
21 experience and knowledge they decided that the usual follow-up would be every 3 to 6  
22 months. Having such checks would ensure that equipment failures are identified early and  
23 that the treatment is effective.

24 They noted that good documentation is important for consistency of care and recommended  
25 that the preferences for future assistive ventilation should be discussed and recorded in an  
26 advance care plan. The objective of an advance care plan is to facilitate people receiving the  
27 medical care that is consistent with their values, goals and preferences.

28 The committee discussed that, when non-invasive ventilation techniques fail to meet agreed  
29 goals, alternative options like tracheostomy or supportive care should be considered. The  
30 committee discussed that, although tracheostomy can be effective in selected cases, it can  
31 be helpful when people do not respond to other ventilation techniques. However, it is an  
32 invasive ventilation technique and is therefore associated with a risk of adverse events.  
33 Respiratory failure is a life limiting condition and people may prefer not to have such invasive  
34 treatments when other treatments have not been effective. They may therefore choose to  
35 have supportive care (care focused on relieving symptoms caused by serious illnesses)  
36 instead. Due to the lack of evidence and an approach that needs to be tailored to each  
37 individual's circumstances the committee did not want to be too prescriptive about this and  
38 did not make a strong recommendation on tracheostomy or any other alternative treatment  
39 option.

40 Due to the lack of evidence the committee discussed whether a research recommendation  
41 should be prioritised. However, they agreed that the treatment with assisted ventilation is  
42 individualised and would vary according to needs, preferences and circumstances. They  
43 therefore decided that it would be difficult to conduct research in this.

#### 44 **Cost effectiveness and resource use**

45 The committee noted that no relevant published economic evaluations had been identified for  
46 this topic.

47 The committee considered that these recommendations would just reinforce current practice.  
48 The committee thought there may be some cost savings from ending assisted ventilation  
49 earlier where it is not being effective but that this resource saving would only be small.

1 **Other factors the committee took into account**

2 The committee were aware of NICE Technology appraisal guidance [TA 139] [Continuous](#)  
3 [positive airway pressure for the treatment of obstructive sleep apnoea/hypopnoea syndrome](#)  
4 and cross referred it for further information on assisted ventilation.

5 Although no evidence exists in the population for cerebral palsy, there is evidence for  
6 assisted ventilation in progressive neuromuscular conditions. When they drafted  
7 recommendations the committee took some of these recommendations into account in their  
8 discussion (for example the NICE guideline NG42 on [motor neuron disease](#)).

9 **References**

10 No studies were included in this review.

11

# 1 Appendices

## 2 Appendix A – Review protocols

3 Review protocol for review question C2: Does assisted ventilation improve quality of life for adults with cerebral palsy who have a chronic  
 4 respiratory disorder (including respiratory failure)?

5 **Table 3: Review protocol for assisted ventilation in adults with cerebral palsy with chronic respiratory disorder**

Field (based on PRISMA-P)	Content
Review question	Does assisted ventilation improve quality of life for adults with cerebral palsy who have a chronic respiratory disorder (including respiratory failure)?
Type of review question	Intervention
Objective of the review	The aim of this review is to determine the effectiveness of the assisted ventilation techniques in adults with cerebral palsy who have a chronic respiratory disorder (including respiratory failure) to improve their quality of life.
Eligibility criteria – population/disease/condition/issue/domain	Adults aged 25 and over with cerebral palsy who have a chronic respiratory disorder (including respiratory failure)
Eligibility criteria – intervention(s)/exposure(s)/prognostic factor(s)	<ul style="list-style-type: none"> <li>• Assisted ventilation techniques:                             <ul style="list-style-type: none"> <li>○ Non-invasive ventilation (NIV)</li> <li>○ Ventilation via tracheostomy</li> </ul> </li> </ul>
Eligibility criteria – comparator(s)/control or reference (gold) standard	<ul style="list-style-type: none"> <li>• No assisted ventilation</li> <li>• NIV versus standard care</li> <li>• NIV versus ventilation via tracheostomy</li> </ul>
Outcomes and prioritisation	<p><b>Critical outcomes</b></p> <ul style="list-style-type: none"> <li>• Hospital admissions</li> <li>• Overall survival</li> <li>• Quality of life (carer or self-reported)</li> </ul> <p><b>Important outcomes</b></p>

Field (based on <u>PRISMA-P</u> )	Content
	<ul style="list-style-type: none"> <li>• Treatment complications</li> <li>• Daytime sleepiness and fatigue</li> </ul> <p>Minimally important differences</p> <ul style="list-style-type: none"> <li>• Any statistically significant improvement in overall survival will be considered clinically important</li> <li>• MID in HrQoL for people with chronic respiratory disease (Wyrwich KW, Tierney WM, Babu AN, Kroenke K, Wolinsky FD. A Comparison of Clinically Important Differences in Health-Related Quality of Life for Patients with Chronic Lung Disease, Asthma, or Heart Disease. Health Services Research. 2005;40(2):577-592).</li> <li>• Other dichotomous outcomes will use default MIDs [RR thresholds of 0.80 and 1.2]</li> <li>• Other continuous outcomes will use default MIDs [0.5 times the SD of the control group]</li> </ul>
Eligibility criteria – study design	<p>Only published full text papers –</p> <ul style="list-style-type: none"> <li>• Systematic reviews of RCTs</li> <li>• RCTs</li> <li>• Comparative cohort studies (only if RCTs unavailable or limited data to inform decision making)</li> <li>• Cross-sectional studies</li> </ul> <p>Consider conference abstract only if related to RCTs.</p>
Other inclusion exclusion criteria	None
Proposed sensitivity/sub-group analysis, or meta-regression	<p>In the presence of heterogeneity, the following subgroups will be considered for sensitivity analysis:</p> <ul style="list-style-type: none"> <li>• Population subgroups: <ul style="list-style-type: none"> <li>○ Level of functional disability</li> <li>○ Severity of chronic respiratory disorder</li> <li>○ Learning difficulties</li> </ul> </li> </ul>

Field (based on <u>PRISMA-P</u> )	Content
	<ul style="list-style-type: none"> <li>• Intervention subgroups:               <ul style="list-style-type: none"> <li>○ Type of assisted ventilation</li> <li>○ Setting (e.g at home, in hospital)</li> <li>○ Age at initiation of ventilator use</li> <li>○ Continuous vs night time</li> </ul> </li> </ul> <p>Severity of chronic respiratory disorder and type of assisted ventilation will be also considered important confounders which ideally should be adjusted for in any included comparative observational studies.</p>
Selection process – duplicate screening/selection/analysis	A random sample of the references identified in the search will be sifted by a second reviewer. This sample size will be 10% of the total, or 100 studies if the search identifies fewer than 1000 studies. All disagreements in study inclusion will be discussed and resolved between the two reviewers. The senior systematic reviewer or guideline lead will be involved if discrepancies cannot be resolved between the two reviewers.
Data management (software)	STAR was used to sift through the references identified by the search
Information sources – databases and dates	Database(s): Embase 1974 to Present, Ovid MEDLINE(R) In-Process & Other Non-Indexed Citations and Ovid MEDLINE(R) 1946 to Present;
Identify if an update	Not an update
Author contacts	For details please see the guideline in development web site.
Highlight if amendment to previous protocol	For details please see section 4.5 of <a href="#">Developing NICE guidelines: the manual 2014</a> .
Search strategy – for one database	For details please see appendix B.
Data collection process – forms/duplicate	A standardised evidence table format will be used, and published as appendix D (clinical evidence tables) or H (economic evidence tables) of the full guideline.
Data items – define all variables to be collected	For details please see evidence tables in appendix D (clinical evidence tables) or H (economic evidence tables).

Field (based on PRISMA-P)	Content
Methods for assessing bias at outcome/study level	Standard study checklists were used to critically appraise individual studies. For details please see section 6.2 of <a href="#">Developing NICE guidelines: the manual 2014</a> . The risk of bias across all available evidence was evaluated for each outcome using an adaptation of the 'Grading of Recommendations Assessment, Development and Evaluation (GRADE) toolbox' developed by the international GRADE working group <a href="http://www.gradeworkinggroup.org/">http://www.gradeworkinggroup.org/</a> .
Criteria for quantitative synthesis	For details please see section 6.4 of <a href="#">Developing NICE guidelines: the manual 2014</a> .
Methods for quantitative analysis – combining studies and exploring (in)consistency	For details please see the methods in supplementary document C.
Meta-bias assessment – publication bias, selective reporting bias	For details please see section 6.2 of <a href="#">Developing NICE guidelines: the manual 2014</a> .
Confidence in cumulative evidence	For details please see sections 6.4 and 9.1 of <a href="#">Developing NICE guidelines: the manual 2014</a> .
Rationale/context – what is known	For details please see the introduction to the evidence review in the full guideline.
Describe contributions of authors and guarantor	A multidisciplinary committee developed the evidence review. The committee was convened by the National Guideline Alliance (NGA) and chaired by Dr Paul Eunson in line with section 3 of <a href="#">Developing NICE guidelines: the manual 2014</a> . Staff from the NGA undertook systematic literature searches, appraised the evidence, conducted meta-analysis and cost effectiveness analysis where appropriate, and drafted the guideline in collaboration with the committee. For details please see the methods in supplementary document C.
Sources of funding/support	NGA is funded by NICE and hosted by the Royal College of Obstetricians and Gynaecologists.
Name of sponsor	NGA is funded by NICE and hosted by the Royal College of Obstetricians and Gynaecologists.
Roles of sponsor	NICE funds NGA to develop guidelines for those working in the NHS, public health and social care in England.

Field (based on <b>PRISMA-P</b> )	Content
PROSPERO registration number	Not applicable

1 *GRADE: Grading of Recommendations Assessment, Development and Evaluation; HrQoL: Health related quality of life; MID: minimal important difference; NGA: National*  
2 *Guideline Alliance; NHS: National Health Service; NICE: National Institute for Health and Care Excellence; NIV: Non-invasive ventilation; RCT: randomised controlled trial;*  
3 *RoB: risk of bias; RR: risk ratio; SD: standard deviation.*

## Appendix B – Literature search strategies

Literature search strategy for review question C2: Does assisted ventilation improve quality of life for adults with cerebral palsy who have a chronic respiratory disorder (including respiratory failure)?

This appendix is a combined search strategy and will be the same for all the evidence reviews for the C review questions as listed below:

C1: What is the most effective protocol for monitoring respiratory health in adults with cerebral palsy?

C2: Does assisted ventilation improve quality of life for adults with cerebral palsy who have a chronic respiratory disorder (including respiratory failure)?

C3: Are prophylactic treatments (for example, antibiotics, chest physiotherapy, cough assistance) effective in preventing respiratory infections in adults with cerebral palsy?

### Database: Medline & Embase (Multifile)

Database(s): Embase 1974 to 2018 March 22, Ovid MEDLINE(R) In-Process & Other Non-Indexed Citations and Ovid MEDLINE(R) 1946 to Present

**Table 4: Last searched on 22 March 2018**

#	Searches
1	exp Cerebral Palsy/ use prmz
2	exp cerebral palsy/ use oomezd
3	((cerebral or brain or central) adj2 (pal* or paraly#s or pares#s)).tw.
4	cerebral palsy.ti,ab.
5	little? disease.tw.
6	((hemipleg* or dipleg* or tripleg* or quadripleg* or unilateral*) adj5 spastic*).tw.
7	((hemipleg* or dipleg* or tripleg* or quadripleg* or unilateral*) adj3 ataxi*).tw.
8	or/1-6
9	limit 8 to english language
10	limit 9 to (adult <18 to 64 years> or aged <65+ years>) use oomezd [Limit not valid in Ovid MEDLINE(R),Ovid MEDLINE(R) In-Process; records were retained]
11	limit 9 to "all adult (19 plus years)" [Limit not valid in Embase; records were retained]
12	11 use prmz
13	or/10,12
14	exp Respiration/ or exp Respiration, Artificial/ or exp Respiratory Insufficiency/ or exp Respiration Disorders/ or exp Respiratory Tract Infections/ or exp Respiratory Aspiration/ or exp Pneumonia, Aspiration/ or exp Respiratory System Abnormalities/ or exp Respiratory Therapy/ or exp Respiratory Function Tests/ or exp Respiratory Paralysis/ or exp Respiratory Mechanics /or exp Positive-Pressure Respiration/ or exp Positive-Pressure Respiration, Intrinsic/ or exp Continuous Positive Airway Pressure/ or exp Respiratory Distress Syndrome, Adult/ or exp Respiratory Sounds/ or exp Gastroesophageal Reflux/ or exp Hypoxia/ or exp Sleep Apnea Syndromes/ or exp Sleep Apnea, Obstructive/ or exp Airway Obstruction/ or exp Tracheotomy/ or exp Bronchial Diseases/ or exp Bronchitis/ or exp Bronchiectasis/ or exp Bronchoscopy/ or exp Laryngoscopy/ or exp

#	Searches
	Tracheobronchomalacia/ or exp Tracheal Diseases/ or exp Adenoidectomy/ or exp Tonsillectomy/ or exp Pulmonary Disease, Chronic Obstructive/ or exp Pulmonary Ventilation/ or exp Airway Management/ or exp Suction/ or exp Airway Resistance/ or exp Cough/ or exp Bronchial Spasm/ or exp Ventilator Weaning/ or exp Ventilators, Mechanical/ or exp Ventilators, Negative-Pressure/ or exp Pulmonary Edema/ or exp Oxygen Consumption/ or exp Oxygen Inhalation Therapy/ or exp Administration, Intranasal/ or exp Catheterization/ or exp Intubation, Intratracheal/ or exp Laryngeal Masks/ or exp Masks/ or exp Catheter Ablation/ or exp Pneumonia/ or exp Pneumonia, Ventilator-Associated/ or exp Ventilator-Induced Lung Injury/ or exp Mouth Breathing/ or exp Phrenic Nerve/ or exp Diaphragm/ or exp Hypoventilation/ or exp Oximetry/ or exp Oxyhemoglobins/ or exp Oxygen/ or exp Carbon Dioxide/ or exp Blood Gas Analysis/ or exp Tidal Volume/ or exp Sleep/ or exp Rest/ or exp Fatigue/ or exp Home Care Services/ or exp Self-Help Devices/ or exp Equipment Failure Analysis/ or exp Intensive Care Units/ or exp Dilatation/ or exp Critical Care/ or exp Self Care/ or exp "Quality of Life"/ or exp Ambulatory Care/ or exp Patient Admission/ or exp Hospitalization/ or exp "Length of Stay"/ or exp Institutionalization/ or exp Physical Therapy Modalities/ or exp Pulmonologists/ or exp Breathing Exercises/ or exp Anti-Bacterial Agents/ or exp Drug Resistance, Bacterial/ or exp Albuterol/ or exp "Nebulizers and Vaporizers"/ or exp Bronchodilator Agents/ or exp Administration, Inhalation/ or exp Saline Solution, Hypertonic/ or exp Influenza Vaccines/ or exp Gastrostomy/ or exp Deglutition Disorders/ or exp Deglutition/ or exp Chest Wall Oscillation/ or exp Asthma/ or exp Bronchopulmonary Dysplasia/ or exp Scoliosis/ or exp Amoxicillin/ or exp Penicillins/ or exp Doxycycline/ or exp Clarithromycin/ or exp Bacterial Infections/co [Complications]
15	14 use prmz
16	exp breathing/ or exp artificial ventilation/ or exp respiratory failure/ or exp breathing disorder/ or exp respiratory tract infection/ or exp acid aspiration/ or exp aspiration pneumonia/ or exp respiratory tract malformation/ or exp respiratory care/ or exp oxygen consumption/ or exp diaphragm paralysis/ or exp positive end expiratory pressure/ or exp adult respiratory distress syndrome/ or exp abnormal respiratory sound/ or exp gastroesophageal reflux/ or exp hypoxia/ or exp sleep disordered breathing/ or exp airway obstruction/ or exp dysphagia/ or exp swallowing/ or exp tracheotomy/ or exp lung functioning test/ or exp bronchus disease/ or exp bronchitis/ or exp bronchiectasis/ or exp bronchoscopy/ or exp laryngoscopy/ or exp tracheobronchomalacia/ or exp trachea disease/ or exp adenoidectomy/ or exp apnea monitoring/ or exp tonsillectomy/ or exp chronic obstructive lung disease/ or exp lung ventilation/ or exp breathing mechanics/ or exp respiration control/ or exp suction drainage/ or exp airway suction device/ or exp suction/ or exp tracheal suction catheter/ or exp suction pump/ or exp airway resistance/ or exp coughing/ or exp bronchospasm/ or exp ventilator/ or exp ventilator weaning/ or exp mechanical ventilator/ or exp lung edema/ or exp oxygen therapy/ or exp intranasal drug administration/ or exp catheterization/ or exp endotracheal intubation/ or exp laryngeal mask/ or exp mask/ or exp catheter ablation/ or exp ventilator associated pneumonia/ or exp pneumonia/ or exp ventilator induced lung injury/ or exp mouth breathing/ or exp phrenic nerve/ or exp diaphragm/ or exp hypoventilation/ or exp oximetry/ or exp oxyhemoglobin/ or exp oxygen/ or exp carbon dioxide/ or exp blood gas analysis/ or exp tidal volume/ or exp rest/ or exp sleep/ or exp sleep disordered breathing/ or exp fatigue/ or exp home care/ or exp self help device/ or exp device failure analysis/ or exp intensive care unit/ or exp dilatation/ or exp intensive care/ or exp self care/ or exp "quality of life"/ or exp ambulatory care/ or exp hospital admission/ or exp hospitalization/ or exp "length of stay"/ or exp institutionalization/ or exp physiotherapy/ or exp pulmonologist/ or exp breathing exercise/ or exp antiinfective agent/ or exp bacterial infection/ or exp antibiotic resistance/ or exp antibiotic agent/ or exp salbutamol/ or exp nebulizer/ or exp vaporizer/ or exp bronchodilating agent/ or exp inhalational drug administration/ or exp sodium chloride/ or exp influenza vaccine/ or exp gastrostomy/ or exp asthma/ or exp lung dysplasia/ or exp

#	Searches
	scoliosis/ or exp amoxicillin/ or exp penicillin derivative/ or exp doxycycline/ or exp clarithromycin/
17	16 use oemezd
18	(respirat* or breath* or ventilat* or tracheo* or trachea* or intratracheal or intubat* or catheter* or airway* or mask* or tent* or sleep apn?ea or tube* or nasotracheal or CNT or obstruct* or mouth* or nose* or nasal or intranasal or nasogastric or failure or distress or pneumon* or lung* or phrenic nerve* or pulmonary* or diaphragm* or tracheo-bronchomalacia or hypoventilat* or positive airway pressure* or negative pressure* or CPAP or negative pressure chamber* or NPC or assist* or manag* or support* or help* or complicat* or leak* or prevent* or prophyla* or monitor* or assistive technology or hypox* or bronch* or bronchopulmonary or laryn* or adenoid* or tonsil* or resistan* or edema* or oxygen* or carbon dioxide or CO2 or inhal* or oximetr* or oxyhemoglobin* or tidal volume* or sleep* or fatigue* or daytime function* or home care or self-help* or self-care* or dilat* or cough* or chest physiotherapy* or antibiot* or critical or quality or hospital admission* or stay or institutional* or thermoplastic patient-ventilator tubing interface* or bedside percutaneous dilatational tracheostomy or PDT or BIPAP or chest infection* or inflammat* or aspiration* or tachypnoea or bronchial spasm* or phlegm* or wheez* or choking or choke* or swallow* or salbutamol or hyperinflation or deglutition* or oscillation* or nebuli?er* or vapor?er* or oral secretion* or saline or oro-pharyngeal suction* or saturation* or vaccine* or pulmonologist* or gastrostom* or bronchitis or percussion* or chest wall vibration* or kyphoscoliosis or amoxicillin or penicillin or doxycycline or clarithromycin).ti,ab.
19	15 or 17 or 18
20	13 and 19
21	conference abstract.pt. use oemezd
22	letter.pt. or LETTER/ use oemezd
23	Letter/ use prmz
24	EDITORIAL/ use prmz
25	editorial.pt. use oemezd
26	NEWS/ use prmz
27	exp HISTORICAL ARTICLE/ use prmz
28	note.pt. use oemezd
29	ANECDOTES AS TOPIC/ use prmz
30	COMMENT/ use prmz
31	CASE REPORT/ use prmz
32	CASE REPORT/ use oemezd
33	CASE STUDY/ use oemezd
34	(letter or comment* or abstracts).ti.
35	or/21-34
36	RANDOMIZED CONTROLLED TRIAL/ use prmz
37	RANDOMIZED CONTROLLED TRIAL/ use oemezd
38	random*.ti,ab.
39	or/36-38
40	35 not 39
41	ANIMALS/ not HUMANS/ use prmz
42	ANIMAL/ not HUMAN/ use oemezd

#	Searches
43	exp ANIMALS, LABORATORY/ use prmz
44	exp ANIMAL EXPERIMENTATION/ use prmz
45	exp MODELS, ANIMAL/ use prmz
46	exp RODENTIA/ use prmz
47	NONHUMAN/ use oomezd
48	exp ANIMAL EXPERIMENT/ use oomezd
49	exp EXPERIMENTAL ANIMAL/ use oomezd
50	ANIMAL MODEL/ use oomezd
51	exp RODENT/ use oomezd
52	(rat or rats or mouse or mice).ti.
53	or/40-52
54	20 not 53

### Database: Cochrane Library

**Table 5: Last searched on 22 March 2018**

ID	Search
#1	MeSH descriptor: [Cerebral Palsy] explode all trees
#2	((cerebral or brain or central) N2 (pal* or paralys?s or pare?s))
#3	((hemipleg* or dipleg* or tripleg* or quadripleg* or unilateral*) N5 spastic*)
#4	((hemipleg* or dipleg* or tripleg* or quadripleg* or unilateral*) N3 ataxi*)
#5	#1 or #2 or #3 or #4
#6	MeSH descriptor: [Respiration] explode all trees
#7	MeSH descriptor: [Respiration, Artificial] explode all trees
#8	MeSH descriptor: [Respiratory Insufficiency] explode all trees
#9	MeSH descriptor: [Respiration Disorders] explode all trees
#10	MeSH descriptor: [Respiratory Tract Infections] explode all trees
#11	MeSH descriptor: [Respiratory Aspiration] explode all trees
#12	MeSH descriptor: [Pneumonia, Aspiration] explode all trees
#13	MeSH descriptor: [Respiratory System Abnormalities] explode all trees
#14	MeSH descriptor: [Respiratory Therapy] explode all trees
#15	MeSH descriptor: [Respiratory Paralysis] explode all trees
#16	MeSH descriptor: [Positive-Pressure Respiration] explode all trees
#17	MeSH descriptor: [Positive-Pressure Respiration, Intrinsic] explode all trees
#18	MeSH descriptor: [Continuous Positive Airway Pressure] explode all trees
#19	MeSH descriptor: [Respiratory Distress Syndrome, Adult] explode all trees
#20	MeSH descriptor: [Respiratory Sounds] explode all trees
#21	MeSH descriptor: [Gastroesophageal Reflux] explode all trees
#22	MeSH descriptor: [Hypoxia] explode all trees
#23	MeSH descriptor: [Sleep Apnea Syndromes] explode all trees
#24	MeSH descriptor: [Sleep Apnea, Obstructive] explode all trees
#25	MeSH descriptor: [Airway Obstruction] explode all trees

ID	Search
#26	MeSH descriptor: [Tracheotomy] explode all trees
#27	MeSH descriptor: [Bronchial Diseases] explode all trees
#28	MeSH descriptor: [Bronchitis] explode all trees
#29	MeSH descriptor: [Bronchiectasis] explode all trees
#30	MeSH descriptor: [Bronchoscopy] explode all trees
#31	MeSH descriptor: [Laryngoscopy] explode all trees
#32	MeSH descriptor: [Tracheobronchomalacia] explode all trees
#33	MeSH descriptor: [Tracheal Diseases] explode all trees
#34	MeSH descriptor: [Adenoidectomy] explode all trees
#35	MeSH descriptor: [Tonsillectomy] explode all trees
#36	MeSH descriptor: [Pulmonary Disease, Chronic Obstructive] explode all trees
#37	MeSH descriptor: [Pulmonary Ventilation] explode all trees
#38	MeSH descriptor: [Pulmonary Ventilation] explode all trees
#39	MeSH descriptor: [Airway Management] explode all trees
#40	MeSH descriptor: [Suction] explode all trees
#41	MeSH descriptor: [Airway Resistance] explode all trees
#42	MeSH descriptor: [Cough] explode all trees
#43	MeSH descriptor: [Bronchial Spasm] explode all trees
#44	MeSH descriptor: [Ventilator Weaning] explode all trees
#45	MeSH descriptor: [Ventilators, Mechanical] explode all trees
#46	MeSH descriptor: [Ventilators, Negative-Pressure] explode all trees
#47	MeSH descriptor: [Pulmonary Edema] explode all trees
#48	MeSH descriptor: [Oxygen Inhalation Therapy] explode all trees
#49	MeSH descriptor: [Administration, Intranasal] explode all trees
#50	MeSH descriptor: [Catheterization] explode all trees
#51	MeSH descriptor: [Intubation, Intratracheal] explode all trees
#52	MeSH descriptor: [Laryngeal Masks] explode all trees
#53	MeSH descriptor: [Masks] explode all trees
#54	MeSH descriptor: [Catheter Ablation] explode all trees
#55	MeSH descriptor: [Pneumonia] explode all trees
#56	MeSH descriptor: [Pneumonia, Ventilator-Associated] explode all trees
#57	MeSH descriptor: [Ventilator-Induced Lung Injury] explode all trees
#58	MeSH descriptor: [Mouth Breathing] explode all trees
#59	MeSH descriptor: [Phrenic Nerve] explode all trees
#60	MeSH descriptor: [Diaphragm] explode all trees
#61	MeSH descriptor: [Hypoventilation] explode all trees
#62	MeSH descriptor: [Oximetry] explode all trees
#63	MeSH descriptor: [Oxyhemoglobins] explode all trees
#64	MeSH descriptor: [Oxygen] explode all trees
#65	MeSH descriptor: [Carbon Dioxide] explode all trees
#66	MeSH descriptor: [Blood Gas Analysis] explode all trees

ID	Search
#67	MeSH descriptor: [Tidal Volume] explode all trees
#68	MeSH descriptor: [Sleep] explode all trees
#69	MeSH descriptor: [Fatigue] explode all trees
#70	MeSH descriptor: [Home Care Services] explode all trees
#71	MeSH descriptor: [Self-Help Devices] explode all trees
#72	MeSH descriptor: [Equipment Failure Analysis] explode all trees
#73	MeSH descriptor: [Intensive Care Units] explode all trees
#74	MeSH descriptor: [Dilatation] explode all trees
#75	MeSH descriptor: [Critical Care] explode all trees
#76	MeSH descriptor: [Self Care] explode all trees
#77	MeSH descriptor: [Quality of Life] explode all trees
#78	MeSH descriptor: [Ambulatory Care] explode all trees
#79	MeSH descriptor: [Patient Admission] explode all trees
#80	MeSH descriptor: [Hospitalization] explode all trees
#81	MeSH descriptor: [Length of Stay] explode all trees
#82	MeSH descriptor: [Institutionalization] explode all trees
#83	MeSH descriptor: [Physical Therapy Modalities] explode all trees
#84	MeSH descriptor: [Pulmonologists] explode all trees
#85	MeSH descriptor: [Breathing Exercises] explode all trees
#86	MeSH descriptor: [Anti-Bacterial Agents] explode all trees
#87	MeSH descriptor: [Drug Resistance, Bacterial] explode all trees
#88	MeSH descriptor: [Albuterol] explode all trees
#89	MeSH descriptor: [Nebulizers and Vaporizers] explode all trees
#90	MeSH descriptor: [Bronchodilator Agents] explode all trees
#91	MeSH descriptor: [Administration, Inhalation] explode all trees
#92	MeSH descriptor: [Saline Solution, Hypertonic] explode all trees
#93	MeSH descriptor: [Influenza Vaccines] explode all trees
#94	MeSH descriptor: [Gastrostomy] explode all trees
#95	MeSH descriptor: [Deglutition Disorders] explode all trees
#96	MeSH descriptor: [Deglutition] explode all trees
#97	MeSH descriptor: [Chest Wall Oscillation] explode all trees
#98	MeSH descriptor: [Asthma] explode all trees
#99	MeSH descriptor: [Bronchopulmonary Dysplasia] explode all trees
#100	MeSH descriptor: [Scoliosis] explode all trees
#101	MeSH descriptor: [Amoxicillin] explode all trees
#102	MeSH descriptor: [Penicillins] explode all trees
#103	MeSH descriptor: [Doxycycline] explode all trees
#104	MeSH descriptor: [Clarithromycin] explode all trees
#105	MeSH descriptor: [Bacterial Infections] explode all trees and with qualifier(s): [Complications - CO]
#106	respirat* or breath* or ventilat* or tracheo* or trachea* or intratracheal or intubat* or catheter* or airway* or mask* or tent* or sleep apn?ea or tube* or nasotracheal or CNT or

ID	Search
	obstruct* or mouth* or nose* or nasal or intranasal or nasogastric or failure or distress or pneumon* or lung* or phrenic nerve* or pulmonary* or diaphragm* or tracheo-bronchomalacia or hypoventilat* or positive airway pressure* or negative pressure* or CPAP or negative pressure chamber* or NPC or assist* or manag* or support* or help* or complicat* or leak* or prevent* or prophyla* or monitor* or assistive technology or hypox* or bronch* or bronchopulmonary or laryn* or adenoid* or tonsil* or resistan* or edema* or oxygen* or carbon dioxide or CO2 or inhal* or oximetr* or oxyhemoglobin* or tidal volume* or sleep* or fatigue* or daytime function* or home care or self-help* or self-care* or dilat* or cough* or chest physiotherapy* or antibiot* or critical or quality or hospital admission* or stay or institutional* or thermoplastic patient-ventilator tubing interface* or bedside percutaneous dilatational tracheostomy or PDT or BIPAP or chest infection* or inflammat* or aspiration* or tachypnoea or bronchial spasm* or phlegm* or wheez* or choking or choke* or swallow* or salbutamol or hyperinflation or deglutition* or oscillation* or nebuli?er* or vaporizer* or oral secretion* or saline or oro-pharyngeal suction* or saturation* or vaccine* or pulmonologist* or gastrostom* or bronchitis or percussion* or chest wall vibration* or kyphoscoliosis or amoxicillin or penicillin or doxycycline or clarithromycin
#107	{or #6-#106}
#108	#5 and #107

#### Database: Web of Science

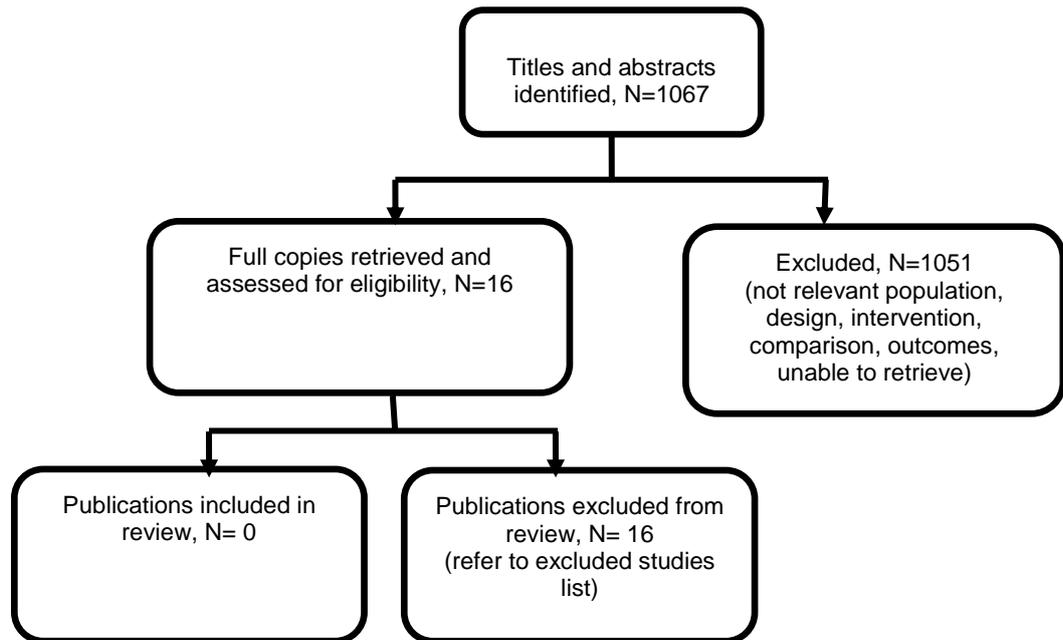
**Table 6: Last searched on 22 March 2018**

#4	(#3) AND LANGUAGE: (English)
#3	#2 AND #1
#2	ts=Artificial Respiration or ts=Respiratory Tract Infection* or ts=Aspiration or ts=respirat* failure* or ts=Pneumoni* or ts=Respiratory Therapy or ts=Respiratory Distress Syndrome or ts=Airway* Obstruction* or ts=Bronch* Disease* or ts=Pulmonary Ventil* or ts=mechanical ventilation* or ts=Breathing Exercise* or ts=antibiotic* or ts=Vaccine* or ts=Bacterial Infection* or ts=breath* or ts=mask* or ts=tent* or ts=sleep apn?ea or ts=tube* or ts=hypoventilat* or ts=positive airway pressure* or ts=negative pressure* or ts=chest infection* or ts=inflammat* or ts=oxygen* or ts=carbon dioxide or ts=CO2 or ts=bronchial spasm* or ts=phlegm* or ts=wheez* or ts=choking or ts=choke* or ts=swallow* or ts=salbutamol or ts=percussion* or ts=chest wall vibration* or ts=scoliosis or ts=amoxicillin or ts=penicillin or ts=doxycycline or ts=clarithromycin
#1	ts=cerebral palsy

## Appendix C – Clinical evidence study selection

Clinical evidence study selection for review question C2: Does assisted ventilation improve quality of life for adults with cerebral palsy who have a chronic respiratory disorder (including respiratory failure)?

Figure 1: Flow diagram of clinical article selection for this review



## **Appendix D – Clinical evidence tables**

Clinical evidence tables for review question C2: Does assisted ventilation improve quality of life for adults with cerebral palsy who have a chronic respiratory disorder (including respiratory failure)?

No clinical studies were identified for this review.

## **Appendix E – Forest plots**

Forest plots for review question C2: Does assisted ventilation improve quality of life for adults with cerebral palsy who have a chronic respiratory disorder (including respiratory failure)?

No forest plots were included in this review.

## **Appendix F – GRADE tables**

GRADE tables for review question C2: Does assisted ventilation improve quality of life for adults with cerebral palsy who have a chronic respiratory disorder (including respiratory failure)?

No GRADE tables were included in this review.

## **Appendix G – Economic evidence study selection**

Economic evidence study selection for review question C2: Does assisted ventilation improve quality of life for adults with cerebral palsy who have a chronic respiratory disorder (including respiratory failure)?

No economic evidence was identified for this review.

## **Appendix H – Economic evidence tables**

Economic evidence tables for review question C2: Does assisted ventilation improve quality of life for adults with cerebral palsy who have a chronic respiratory disorder (including respiratory failure)?

No economic evidence was identified for this review.

## **Appendix I – Health economic evidence profiles**

Health economic evidence profiles for review question C2: Does assisted ventilation improve quality of life for adults with cerebral palsy who have a chronic respiratory disorder (including respiratory failure)?

No economic evidence was identified for this review.

## **Appendix J – Health economic analysis**

Health economic analysis for review question C2: Does assisted ventilation improve quality of life for adults with cerebral palsy who have a chronic respiratory disorder (including respiratory failure)?

No economic analysis was included in this review.

## Appendix K – Excluded studies

Clinical and economic lists of excluded studies for review question C2: Does assisted ventilation improve quality of life for adults with cerebral palsy who have a chronic respiratory disorder (including respiratory failure)?

### Clinical studies

**Table 7: Excluded clinical studies for assisted ventilation in adults with cerebral palsy with chronic respiratory disorder**

<b>Excluded studies – C2: Does assisted ventilation improve quality of life for adults with cerebral palsy who have a chronic respiratory disorder (including respiratory failure)?</b>	
<b>Study</b>	<b>Reason for Exclusion</b>
Bischof, F., Hyperbaric oxygen therapy in cerebral palsy, South African Medical Journal, 91, 182, 2001	Letter to editor about a study on children with cerebral palsy.
Chong, H. S., Alfonso, M. R. P., Kim, J. S., Lee, H. M., Moon, S. H., Suk, K. S., Kim, H. S., Usefulness of noninvasive positive-pressure ventilation during surgery of flaccid neuromuscular scoliosis, Journal of Spinal Disorders and Techniques, 28, 298-300, 2015	Mean age of subjects is less than 18 years. Only two subjects with cerebral palsy included.
Heuser, G., Uszler, J. M., Hyperbaric oxygenation for cerebral palsy, Lancet, 357, 2053-2054, 2001	Reports on a study on children with cerebral palsy.
Holland, L.J., Bhambhani, Y.N., Ferrara, M.S., Steadward, R.D., Reliability of the maximal aerobic power and ventilatory threshold in adults with cerebral palsy, Archives of Physical Medicine and Rehabilitation, 75, 687-691, 1994	Not related to assisted ventilation.
James, P. B., Hyperbaric oxygenation for cerebral palsy, Lancet, 357, 2052-2053, 2001	Related to use of hyperbaric oxygen therapy for children.
Kirby, N. A., Barnerias, M. J., Siebens, A. A., An evaluation of assisted cough in quadriparetic patients, Archives of Physical Medicine & Rehabilitation Arch Phys Med Rehabil, 47, 705-10, 1966	Not related to assisted ventilation technique.
Lampe, R., Blumenstein, T., Turova, V., Alves-Pinto, A., Lung vital capacity and oxygen saturation in adults with cerebral palsy, Patient Preference and Adherence, 8, 1691-1697, 2014	Not related to assisted ventilation techniques.
Ntoumenopoulos, G., Berry, M., Camporota, L., Effects of manually-assisted cough combined with postural drainage, saline instillation and airway suctioning in critically-ill patients during high-frequency oscillatory ventilation: a prospective observational single centre trial, Physiotherapy Theory & Practice Physiother, 30, 306-11, 2014	Does not report on subjects with cerebral palsy.
Pareek, N., Williams, J., Hanna, D., Johnson, W.D., Minocha, A., Abell, T.L., Prokinetic therapy reduces aspiration pneumonia in tube-fed patients with severe developmental disabilities, American Journal of Mental Retardation, 112, 467-471, 2007	Does not report on evaluation of assisted ventilation.
Rashid, A., Raj, B., Stoddart, A., Repeat percutaneous dilatational tracheostomy in an awake and unintubated patient, Acta Anaesthesiologica Scandinavica, 51, 378-379, 2007	Case report of a single patient.
Rose, L., McKim, D., Katz, S., Leasa, D., Nonoyama, M., Pedersen, C., Avendano, M., Goldstein, R., Institutional care for long-term	Data on long term mechanical ventilation cases. Cerebral palsy

mechanical ventilation in Canada: A national survey, Canadian Respiratory Journal, 21, 357-362, 2014	cases included in 'others' category with one included subject.
Rose, Louise, Adhikari, Neill Kj, Leasa, David, Fergusson, Dean A, McKim, Douglas, Cough augmentation techniques for extubation or weaning critically ill patients from mechanical ventilation, Cochrane Database of Systematic Reviews, 2017	Not related to evaluation of assisted ventilation techniques.
Shah, R. K., Lander, L., Berry, J. G., Nussenbaum, B., Merati, A., Roberson, D. W., Tracheotomy outcomes and complications: A national perspective, Laryngoscope, 122, 25-29, 2012	Cerebral palsy adults constitute 0.3% of the sample. Tracheostomy cases included both temporary and permanent tracheostomies, and study population was not restricted to those having chronic respiratory conditions.
Vianello, A., Corrado, A., Arcaro, G., Gallan, F., Ori, C., Minuzzo, M., Bevilacqua, M., Mechanical insufflation-exsufflation improves outcomes for neuromuscular disease patients with respiratory tract infections, American journal of physical medicine & rehabilitation, 84, 83-8; discussion 89-91, 2005	Does not report on subjects with cerebral palsy.
vonScheele, B. H. C., Frojd, J. K., Gahn, E. E., Analysis of breathing behavior in cerebral palsy athletes (CPA): Capnography during deep and "normal" breathing, Biofeedback and Self-Regulation, 21, 385-385, 1996	Does not report on evaluation of assisted ventilation techniques.
Yeow, M. E., Your own sweet time: discontinuing ventilator support at home, Journal of Palliative Medicine, 18, 388-9, 2015	Does not report on evaluation of assisted ventilation techniques.

## Economic studies

No economic evidence was identified for this review.

## Appendix L – Research recommendations

Research recommendations for review question C2: Does assisted ventilation improve quality of life for adults with cerebral palsy who have a chronic respiratory disorder (including respiratory failure)?

No research recommendations were made for this review.