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Narrative review

Management of respiratory problems in people with neurodegenerative conditions: a narrative review

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Abstract

Background Respiratory failure and dysfunction are common problems in many neurodegenerative conditions. Although physiotherapists manage these problems, it is not known which treatments have been studied and their efficacy.

Objective To review evidence on the management of respiratory problems in people with neurodegenerative conditions using the PRISMA approach.

Data sources Comprehensive searches were conducted using the following electronic databases from inception to May 2010: HUGEnet, SIGLE, British Library Direct, CINAHL, Medline, AMED and Web of Knowledge. Bibliographies of all studies and systematic reviews were searched by hand.

Study selection Studies were selected based on: self-ventilating participants with neurodegenerative conditions; interventions aimed at improving respiratory function; and any valid and reliable measures of respiratory function as outcomes.

Study appraisal Studies were appraised by one reviewer using the Critical Appraisal Skills Programme. Data were synthesised using a narrative approach.

Results Thirty-five studies were included in the review. The strongest evidence was for the use of non-invasive ventilation for people with amyotrophic lateral sclerosis, although this was weak. The evidence for the use of respiratory muscle training and methods to increase peak cough flow showed a positive effect, but was also weak.

Conclusion There is weak evidence for the positive effects of physiotherapeutic interventions for respiratory problems in people with neurodegenerative conditions. Further work is necessary in specific neurodegenerative conditions to identify why respiratory problems occur, and larger scale studies should be undertaken to investigate management of these problems.

Keywords: Neurodegenerative conditions; Respiratory insufficiency; Physiotherapy

Introduction

Rationale

Respiratory dysfunction is common in neurodegenerative conditions, such as multiple sclerosis [1], amyotrophic lateral sclerosis (ALS) [2] and Huntington's disease [3]. Physiotherapeutic management of respiratory problems is often supportive rather than preventative, only taking place in the middle and late stages of the condition [4]. With the exception of national guidelines for the use of non-invasive ventilation in patients with motor neurone disease [5], there are no national guidelines for the management of respiratory problems in people with Parkinson's disease, Huntington's disease or multiple sclerosis. The British Thoracic Society/Association of Chartered Physiotherapists in Respiratory Care guidelines for the adult, spontaneously breathing patient [6] focus on people with neuromuscular disease but do not provide sufficient detail for neurodegenerative conditions. Neurodegenerative conditions differ from neuromuscular diseases in that the former refers to central neurological disorders, and the latter refers to post-neuromuscular junction disorders. Multiple sclerosis, Parkinson's disease, Huntington's disease and ALS/motor neurone disease are neurodegenerative conditions with central nervous system processing problems and peripheral weakness.

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People with neurodegenerative conditions have difficulty clearing secretions for a number of reasons, including respiratory muscle weakness and bulbar insufficiency [7]. Ineffective gas exchange may occur due to retained secretions, compounded by respiratory muscle weakness influencing the effectiveness of cough. Decreased inspiratory muscle strength may lead to alveolar hypoventilation, ventilation–perfusion mismatch, and further respiratory muscle fatigue due to altered biomechanics [8]. There is a gap in our knowledge about the physiotherapeutic management of respiratory problems in people with neurodegenerative conditions, despite the fact that respiratory problems are the leading cause of death in this population [7].

Objective

The aim of this paper was to review evidence on the management of respiratory problems in people with neurodegenerative conditions using the PRISMA statement [9].

Methods

Search process

A population, intervention, comparison and outcome (PICO) approach was used [10,11]. The population was defined as people with neurodegenerative conditions. The intervention was any physiotherapy-based intervention influencing the respiratory system. No set comparisons were made or follow-up times set. Outcome was any reliable and valid measure of respiratory function, and not respiratory failure alone.

Comprehensive searches were conducted using the following electronic databases from inception to May 2010 (number of studies identified in brackets): HUGEnet (161), SIGLE (624), British Library Direct (192), CINAHL (130), Medline, EMBASE and AMED (4307). Bibliographies of all studies and systematic reviews were searched by hand. Keywords were structured using PICO. Population keywords included 'neuro*', 'Parkinson's disease', 'amyotrophic lateral sclerosis', 'motor neurone disease', 'multiple sclerosis' and 'Huntington's disease'. Intervention keywords included 'physiotherapy' and 'respiratory', and outcome keywords included 'lung'. Subsequent to the initial search and analysis of the categories of evidence found, two further search terms were used: 'respiratory muscle strength' and 'retained secretions'. Box 1 shows the search strategy used in CINAHL, Medline, AMED and EMBASE databases.

Box 1: Search strategy.

1 = physiother*
2 = neuro*
3 = respir*
4 = lung
5 = respiratory muscle strength
6 = retained secretions
7 = 3 OR 4 OR 5 OR 6
8 = 1 AND 2 AND 7
9 = motor neurone disease
10 = amyotrophic lateral sclerosis
11 = multiple sclerosis
12 = Huntington's disease
13 = Parkinsonism disease
14 = 7 OR 8 OR 9 OR 10 OR 11
15 = 12 AND 7

Eligibility criteria, identification and selection of studies

Full-text, English-language randomised controlled trials, experimental studies, and prospective and retrospective observational studies which investigated changes in respiratory function following a physiotherapy-based intervention were included. One reviewer identified and reviewed all titles and abstracts followed by the full text of each article. Exclusion criteria were: the population consisted entirely of patients with neuromuscular conditions such as myesthenia gravis and muscular dystrophies, all members of the population were aged <18 years, the subjects were not breathing spontaneously, the intervention did not influence respiratory function, only one subject was studied, or respiratory failure was the sole outcome measure.

Critical appraisal

Critical appraisal was carried out by one reviewer using the Critical Appraisal Skills Programme (CASP) appraisal tool [12].

Data analysis

Analysis was completed by one reviewer. Due to heterogeneity of populations, interventions and outcome measures, it was not possible to carry out a meta-analysis. A narrative review of all included studies was undertaken.

Results

Study selection

In total, 5414 studies were retrieved; 5368 studies were excluded by the title, abstract or method (Fig. 1), and 11 studies were excluded by the full text (available from authors on request). Descriptive analysis of the remaining 35 studies highlighted three main themes: the problem of retained secretions, the problem of decreased muscle strength, and the influence of exercise on respiratory function. Studies were grouped into these themes for the narrative review.