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Trends in survival from muscular dystrophy in England and Wales and impact on respiratory services

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Summary Respiratory failure is an important terminal event in muscular dystrophy, but increasingly is effectively treated by non-invasive ventilation. This study was designed to assess mortality statistics in this patient group in order to get an indication of future demand.

Mortality data for all deaths from muscular dystrophy registered by death certification in England and Wales between 1993 and 1999 were analysed.

In total, 817 deaths from muscular dystrophy were registered between 1993 and 1999. Annual number of deaths was unchanged over this period. Median age at death (interquartile range) for all cause muscular dystrophy increased from 20 (17–42.5) years in 1993, to 26 (17.5–63) years in 1999. Respiratory failure was the primary or contributory cause of death in 82% of cases. Two thirds of these deaths were during acute infection.

We can expect 100 patients with muscular dystrophy to develop respiratory failure in England and Wales each year, so non-invasive ventilation services probably need to be able to provide for 0.2 new patients per 100,000 population annually. Respiratory services also need to provide adequate monitoring and early treatment of infection in these patients.

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Introduction

Respiratory failure, often exacerbated by respiratory infection, is an important cause of death in patients with muscular dystrophy. Since the early 1990s there has been a significant rise in the use of non-invasive ventilation (NIV) in this population, as it has become widely accepted as an effective means of managing respiratory failure. NIV has subsequently resulted in substantial improvements in survival in patients with slowly progressive muscular dystrophy.[1–3] Even in more rapidly progressive conditions such as Duchenne muscular dystrophy (DMD), NIV has been shown to prolong life by several years.[4] As a consequence, the number of patients with muscular dystrophy using NIV at home is steadily increasing.[5,6] Since respiratory decompensation often occurs at the crucial time when patients are in the process of moving from paediatric to adult services, patients with muscular dystrophy are increasingly being referred to adult respiratory services for respiratory support and consideration of NIV.

In order to provide an adequate service for patients with muscular dystrophy, we need some indication of their future needs and the likely impact on respiratory services, and in particular NIV services, by this population. However, there is very little epidemiological data in the current literature on which to base any assessment of service requirements. The aim of this study is to collect detailed information on the pattern of number of deaths and age at death over time and on cause of death. We have therefore examined mortality statistics for all causes of muscular dystrophy in England and Wales over a 7-year period. This information can then be used to estimate future demand for respiratory services in this population.

Methods

The time trend in age at death and the effects of diagnostic category of muscular dystrophy and gender have been examined. We have also looked for any geographical variation in age at death that might reflect the presence of specialist clinics for the provision of NIV, and have examined the effects of social class as a possible marker of access to and uptake of NIV services.

Data collection

Mortality data for all deaths with muscular dystrophy mentioned on the death certificate were obtained from the National Statistics Office for the period 1993–1999. The data included the underlying cause of death, age at death, place of death, gender, regional health authority of usual residence, and occupational social class.

Underlying cause of death has been documented as text as supplied on the death certificate under Sections 1a, 1b, and 1c and classified as respiratory, cardiac, cardio-respiratory, muscular dystrophy or other. Age at death was recorded as age in years at the last birthday. Five diagnostic categories of muscular dystrophy were identified for the purpose of analysis—DMD, muscular dystrophy (unspecified), Beckers muscular dystrophy (BMD), limb girdle or facioscapulohumoral muscular dystrophy (LG/FSH), congenital muscular dystrophy and other muscular dystrophy (oropharyngeal, neuroaxonal, pseudohypertrophic, Emery Dreifess, Barths type, myotonic, scapuloperoneal, familial, merosine positive, spinal, ischemic, autosomal recessive).

Coding for regional health authorities corresponds to their reorganisation in 1996. Social class included an occupational code for the deceased if aged over 16 or the mother's occupation if under 16, and, respectively, for the spouse or father. Categories are professional and intermediate, skilled non-manual, skilled manual, semi-skilled, unskilled, and other/unknown. For the purpose of analysis the higher occupational social class was used.

Statistical analysis

The data was analysed in terms of median age at death by calendar year in relation to diagnostic category of muscular dystrophy and gender. Descriptive analyses were carried out using Microsoft Excel.

The independent effects of gender, diagnosis, social class, and health authority region of usual residence on the odds of death above the median age for the year of death were estimated by multiple logistic regression (effects were examined in the three most common diagnostic categories) [SPSS version 10].

Results

A total of 817 people (677 males) were registered as having died from muscular dystrophy in England and Wales between 1993 and 1999. Table 1 shows number of deaths each year by diagnostic category and gender.