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Title: Observation cohort study of cause of death in patients with Duchenne muscular dystrophy (DMD)

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Body: Prior to the introduction of home mechanical ventilation (HMV), the major cause of death in patients with Duchenne muscular dystrophy (DMD) was respiratory failure. We performed an observational mortality cohort study of our DMD population over a 6 year period. As a large specialist centre we have a current population of 117 adult patients with DMD and 12.7% of our total patient population died in the 6 year period.

Table to show cause and location of death in patients with DMD over a 6 year period

| n =17 | 2006 | 2007 | 2008 | 2009 | 2010 | 2011 | Total |
|-------------------------------|------|------|------|------|------|------|------------|
| Total Number of Deaths | 1 | 1 | 2 | 5 | 5 | 3 | 17 |
| Total on Ventilators | 1 | 0 | 2 | 3 | 4 | 3 | 13 (76.5%) |
| Death in Hospital | 0 | 1 | 1 | 2 | 5 | 1 | 10 (58.8%) |
| Death out of Hospital | 1 | 0 | 1 | 3 | 0 | 2 | 7 (41.1%) |
| Respiratory Failure | 1 | 0 | 1 | 3 | 0 | 2 | 6 (35.2%) |
| Cardiac Failure | 0 | 1 | 1 | 2 | 5 | 0 | 10 (58.8%) |
| Other Medical | 0 | 0 | 0 | 0 | 0 | 1 | 1 (5.9%) |

Data are expressed in absolute values (%)

Out of a total of 17 deaths in a six year period, only 35.2% died of respiratory failure with a mean age of 22.7 years, despite 76.5% of the patients being dependent on NIV. The major cause of death in this cohort was cardiac failure, with 58.8% of patients dying from cardiac complications at a mean age of 19.1 years.

Cardiac death was only anticipated in one of the 10 cases, and this patient died in the home environment. These data support the need for early regular cardiac surveillance. In addition to the standard electrocardiographic and echocardiographic measurements, the use of extended electrocardiographic measurements may be required. This is supported by the current UK guidelines endorsing yearly screening for patients with DMD once over the age of 10 years. This project has been supported by the Guy's & St Thomas' Charity (Project Code: G080109).