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What is the mortality of Juvenile Dermatomyositis (JDM) in the modern treatment era

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Background

Despite modern treatment approaches Juvenile Dermatomyositis (JDM) remains a serious and potentially life threatening disease. There are few studies which have documented mortality among large series of cases of JDM treated with modern therapeutic approaches. The Juvenile Dermatomyositis National Registry and Repository, UK and Ireland (JDRR) was established in 2000 to facilitate research and improve knowledge about JDM.

Methods

Children were recruited through the JDRR cohort study. Mortality attributed to JDM or its complications was recorded. A survey of contributors in The Juvenile Dermatomyositis Research Group was also conducted to establish mortality in JDM cases that had not been recruited to the JDRR study.

Results

245 children (166 females) with myositis have been recruited to the JDRR. Of these, 208 have a diagnosis of JDM or JDM with overlap features (148 females). The total years of JDM disease documented is 1353 patient years. There have been 2 recorded deaths, a rate of 0.96% or 0.15 per 100 patient years of disease. However physicians in contributing centres were aware of 1 death attributable to JDM in cases that could not be recruited to the study before death.

Conclusion

Mortality due to rare diseases can be difficult to estimate accurately. Within the UK and Ireland JDRR study, mortality due to JDM since 2000 has been 0.96%. However this may underestimate deaths as the JDRR does not necessarily recruit all cases of JDM within the UK. JDM remains a serious a life threatening disease of children despite modern therapies and specialist care.

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