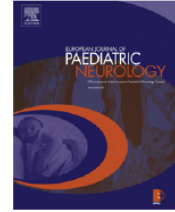




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Original article

Do females with juvenile ceroid lipofuscinosis (Batten disease) have a more severe disease course? The Danish experience

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ABSTRACT

Background: Juvenile neuronal ceroid lipofuscinosis (JNCL; Batten disease) is characterized by progressive visual failure starting at 4–7 years of age, followed by seizures, dementia as well as a progressive decline in motor function. The patients are typically bedridden in the late teens and death usually occurs in the third decade of life. It has been suggested, that females may have a more precipitous decline than do males.

Objective: To compare sex differences in loss of skills and age at death in an unselected population of Danish Adolescents with Batten disease.

Method: Review of hospital records of all 35 Danish patients with JNCL born in the period 1971–2003. The records contain a continuously maintained history of the clinical course and first moments for different events, thus eliminating recall bias.

Results: We found that females with JNCL experienced a later age at diagnosis, but showed an earlier loss of independent functions, and died at an earlier age.

Conclusion: Females with JNCL have a more precipitous decline than males, and die at an earlier age. Further studies are needed in order to provide possible explanations for this difference.

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