



## 16- Overview of advances in educational and social supports for young persons with NCL disorders

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### Highlights

- Children with juvenile-onset Batten disease experience dementia, which results in a plateau in, and eventual loss of cognitive skills over time.
- The educational model for children with JNCL should anticipate cognitive decline through a model of proactive and hastened instruction.
- Children with JNCL should learn skills, whilst still able to, that will support independence for as long as possible.
- Ensuring participation in real life activities is of special importance in adult life for individuals with JNCL.
- Interests/participation can be sustained by interaction with others, environmental adaptations, technical aids, and music activities.

## Abstract

Vision loss, dementia, and motor and speech declines all impact the educational experience of individuals with Batten disease and can adversely impact effective learning. There are as yet limited data to support evidence-based approaches to meeting the educational needs of affected individuals. This paper provides an overview of recent work to evaluate and address educational issues with a life-long perspective relevant to individuals with juvenile-onset neuronal ceroid lipofuscinosis (JNCL) and the professionals that provide them with educational support. In particular, several main activities of the recently completed 'JNCL and Education' project are summarised, including a survey of parents, educational professionals and social/health workers, development of a formative assessment tool to identify and respond to an individual student's strengths and needs in the learning environment, and proposed strategies for prolonging literacy and language skills. A key concept that should be emphasised in the educational plan for students with JNCL is that of 'proactive' and 'hastened' learning, that is, providing an early emphasis on adaptive skills that will be required in the later stages of disease progression when new learning will be more difficult to achieve. An additional key concept is participation in real-life activities to maintain skills and quality of life, particularly in the later stages of disease progression.