



Deafblindness

Theory-of-mind, cognitive functioning and social network in
Alström syndrome

av

Hans-Erik Frölander

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Göteborg

Örebro universitet
Institutionen för hälsovetenskaper
701 82 ÖREBRO

Abstract

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This thesis addresses young adults with Alström syndrome (AS). AS causes acquired deafblindness, a severe, progressive, combined auditory and visual impairment affecting daily life and self-reliance to a degree that full participation depends on help from others and society. AS is an autosomal, recessively inherited single-gene disorder that affects the ALMS1 gene. AS has a multi-systemic pathology including a high incidence of additional multiple endocrine abnormalities, cardiomyopathy, pulmonary fibrosis, restrictive lung disease and progressive hepatic and renal failure leading to reduced life expectancy. The focus in the present thesis is on the development of Theory-of-mind (ToM) and on how ToM relates to the development of certain cognitive skills and the characteristics of the individual social network. ToM refers to the ability to understand the thoughts and feelings of others.

The results reveal that individuals with AS displayed a significantly higher degree of heterogeneity in the performance of ToM tasks, and some individuals with AS performed on an equal level with nondisabled individuals. ToM performance was predicted by verbal ability and executive functioning (EF), whereas working memory capacity (WM) proved to be an indirect predictor. Later onset of visual loss further characterized AS individuals with better ToM. The sizes of the social networks of individuals with AS were smaller relative to those of nondisabled individuals, and many of the acquaintances were professionals working with individuals with AS. The number of friends correlated with ToM performance.

Methods to improve verbal ability and EF, and interventions to enhance social participation in childhood of individuals with AS might prove to be fruitful in addition to assistive technology to establish and maintain friendships in adulthood.

Keywords: Alström syndrome, Deafblindness, Theory-of-mind, Communication, Verbal ability, Executive functions, Working memory, Health problems, Friendship, Social network

Örebro University, SE-701 82 Örebro, Sweden, hans-erik.frolander@spsm.se