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# An exploratory natural history of ataxia of Charlevoix-Saguenay

## A 2-year follow-up

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## Abstract

**Objective** To document the decline of upper and lower limb functions, mobility, and independence in daily living activities in adults with autosomal recessive spastic ataxia of Charlevoix-Saguenay (ARSACS) over a 2-year period.

**Methods** An exploratory longitudinal design was used. Nineteen participants were assessed on 2 occasions 2 years apart. Assessments included the Standardized Finger Nose Test, Nine-Hole Peg Test, Lower Extremity Motor Coordination Test, Berg Balance Scale, 10-m walk test (10mWT), 6-minute walk test (6MWT), Scale for the Assessment and Rating of Ataxia (SARA), and Barthel Index.

**Results** A significant decline was observed between baseline and follow-up for lower limb coordination, balance, walking abilities (10mWT and 6MWT), and overall disease severity

(SARA). All differences were beyond measurement error documented in ARSACS. Results showed no significant decline for upper limb coordination and fine dexterity performance.

**Conclusion** Although ARSACS is a slow, progressive disease, results showed that mobility, balance, and lower limb performance significantly decreased over the 2-year period and that selected outcome measures were able to capture this decline beyond measurement errors.

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