Swallowing & speech deficits are detectable at the pre-ataxic stage of Spinocerebellar Ataxia Type 2

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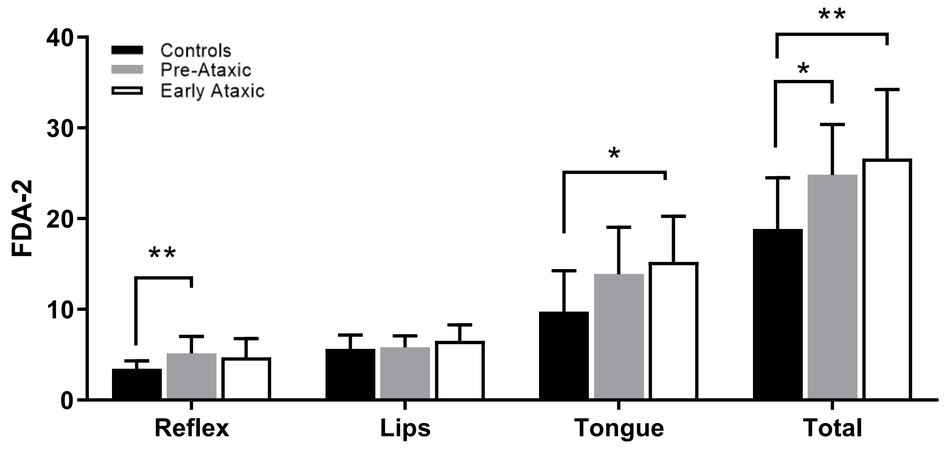
Changes in swallowing and speech function appear to be subtle markers of pre-ataxic and early disease progression in SCA2

BACKGROUND & AIM

- Swallowing and speech difficulties are common in spinocerebellar ataxias (SCA).
- The nature and severity of SCA genotype-specific deficits are not well described.
- More information is needed to characterize swallowing and speech difficulties particularly evolution from **pre-ataxic** to **early-manifest** stages.
- We present a comprehensive study of speech and swallowing function in **pre-ataxic** to **early-stage ataxic** SCA2 individuals, using objective measures of speech combined with detailed measures of swallowing and quality of life.

ORAL MOTOR AND SWALLOWING FUNCTION IN SCA2



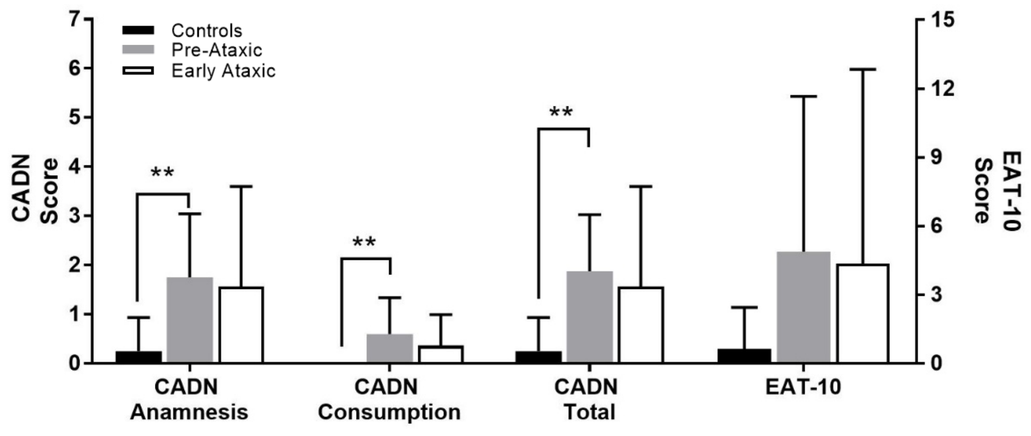


Figure 1. Oral motor and swallowing function in pre-ataxic and early-ataxic SCA2.
*P < 0.05, **P < 0.01, ***P < 0.001. Values represent Mean ± SD.
Oral motor function was assessed using the Frenchay Dysarthria Assessment-2. Swallowing function and associated quality of life was measured through use of the Clinical assessment of dysphagia in neurodegeneration (CADN) and Eating assessment Tool (EAT-10)

ACOUSTIC MEASURES OF SPEECH IN SCA2

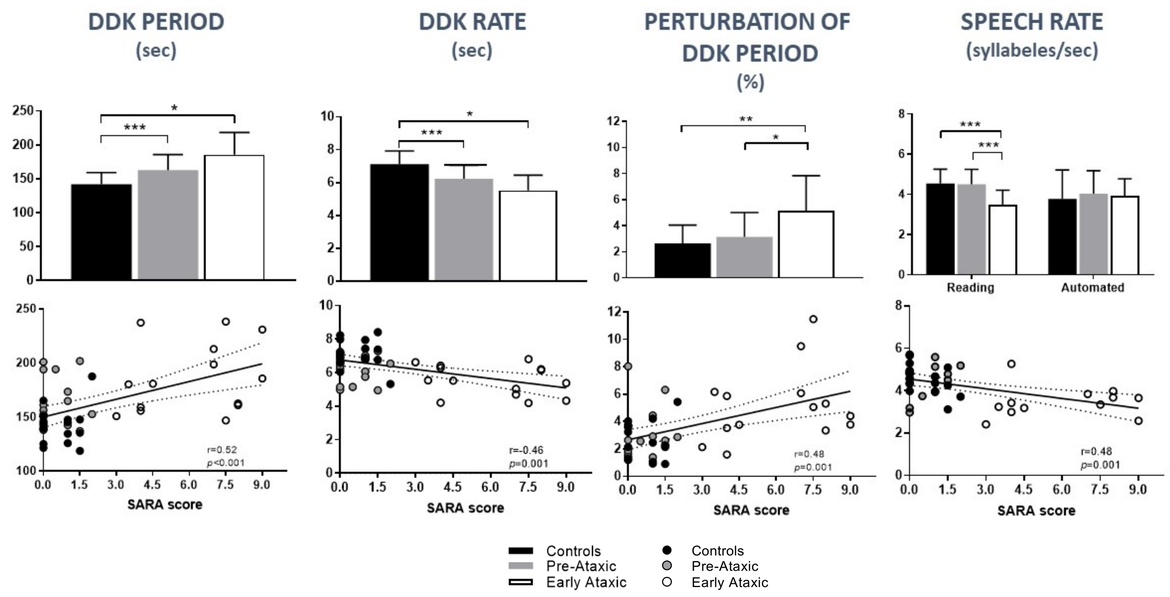


Figure 2. Acoustic measures of syllable repetition and speech timing, and their relationship with SCA2 ataxia severity as measured by SARA.
*P < 0.05, **P < 0.01, ***P < 0.001. Values represent Mean ± SD. Black dotted lines represent 95% Confidence Intervals.

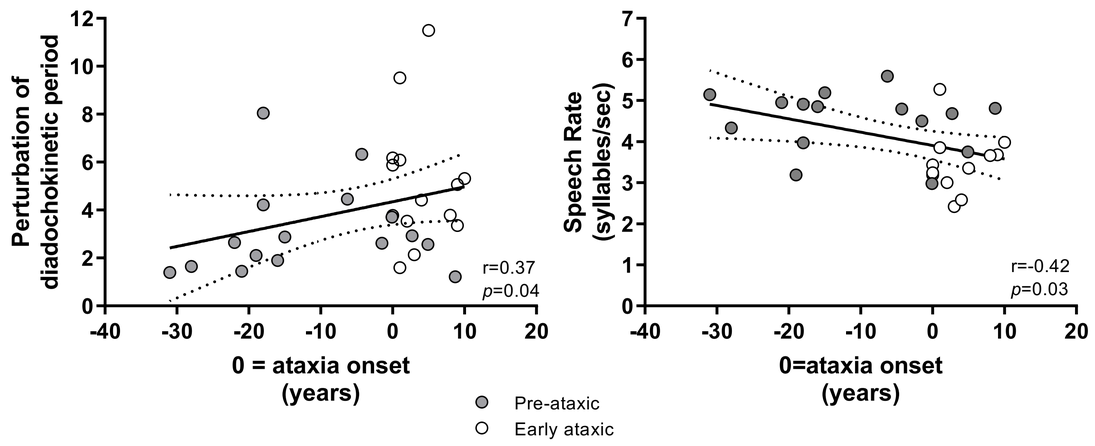
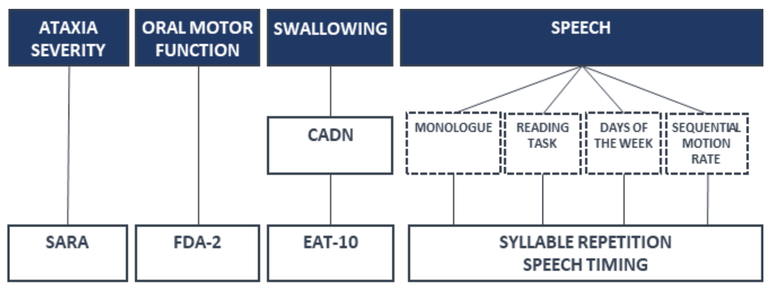


Figure 3. Acoustic measures of syllable repetition and speech timing, and their relationship with onset of ataxia. Dotted lines represent 95% Confidence Intervals.

METHODS

- Forty-six individuals were recruited from the Centre for Research and Rehabilitation in Hereditary Ataxias (CIRAH) in Holguín, Cuba (Table 1).
- Participants underwent a comprehensive battery of assessments including clinician derived ratings of speech function and swallowing, quality of life assessments of swallowing and objective acoustic analysis (Figure 4).

Figure 4. Protocol Schematic



Abbreviations- SARA: Scale for the Assessment and Rating of Ataxia[1], FDA-2: Frenchay Dysarthria Assessment-2[2], CADN: Clinical Assessment of Dysphagia in Neurodegeneration[3], EAT-10: Eating Assessment Tool[4].

	Healthy Controls	Pre-Ataxic SCA2	Early Ataxic SCA2	P
N	16	16	14	
Age (years)	38.38 ± 10.46	39.00 ± 8.86	37.79 ± 10.62	NS
Range	18-59	18-52	18-59	
Sex (n)	7M	5M	7M	NS
	9F	11F	7F	
Age at ataxia onset (years)	-	-	34.00 ± 8.96	NS
Range	-	-	17-45	NS
Ataxia severity (SARA)	0.68 ± 0.60	0.59 ± 0.69	6.14 ± 2.18*	0.001
Range	0-2	0-2	3-9	

Table 1. Participant Characteristics
*P < 0.05 Control vs. Early Ataxic SCA2.

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CADN