Quality of life of Brazilian patients with hereditary angioedema upon multidisciplinary management

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Rationale

We investigated whether a comprehensive multidisciplinary intervention, addressing psychosocial aspects, adherence and assurance among HAE patients would have an added value to medical treatment in improvement quality of life.

Methods

Thirty-three patients with HAE, belonging to a single family with mutation c.351delC in SERPING1, participate in a 14-month intervention. The program was coordinated by an allergist, with additional care provided by a psychiatrist, psychologist, nurse, social worker and nurse technician. The impact of the program was assessed through questionnaires administered at the beginning, and at 8 and 14 months of the intervention. Quality of life was assessed by the Hereditary Angioedema Quality of Life (HAE-QoL) questionnaire, and by SF-36 and Pediatrics Quality of Life (PedsQL). Patients were also evaluated by Depression, Anxiety and Stress (DASS) questionnaire, Hospital Anxiety and Depression Scale (HADS),Beck Depression Inventory (BDI), Beck Anxiety Inventory (BAI), Children's Depression Inventory (CDI), and Child Stress Symptoms Inventory (ISS). The Work Productivity and Activity Impairment Questionnaire: General Health (WPAI-GH) was applied to 20 patients employed during the study.

Results

Table 1. Demographic and clinical characteristics of the study participants.

Parameter		Patients n=33
Age median, range (years)		31 (0.5-80)
Age at onset of symptoms median, range (years)		11 (0.2-49)
Gender	F(%)	18 (54.5%)
	M(%)	15 (45.5%)
Severity score by Ferraro et al [2]		
	Asymptomatic	4 (12.1%)
	Mild	5(14.7%)
	Moderate	8 (24.2%)
	Severe	16 (48.5%)
Severity score by Bygum et al. median (range) [4]		5.0 (0-10)
Patients under long term treatment (%)		23 (69.7%)
Attenuated androgens		17 (74.0%)
Anti-fibrinolytic (tranexamic acid)		5 (21.7%)
pdC1-INH concentrate		1 (4.3%)

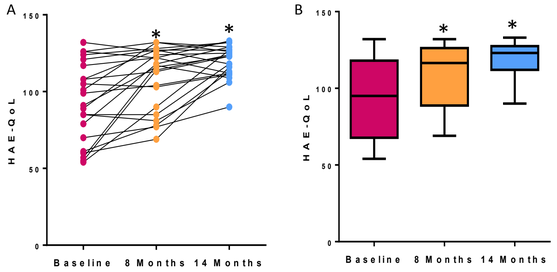

Figure 1. Significant improvement in quality of life was observed using HAE-QoL questionnaire at 8 and 14 months of intervention. Significant increases in mean total HAE-QoL scores of 15.21 (95% Credibility Interval CI 1.23 to 29.77) and 26 (95% CI 14.56 to 39.02) were observed at 8 and 14 months within the study, as compared to baseline (adjusted Bayesian models of beta-binomial regression analysis using OpenBUGS). Data available for 22 adult patients.

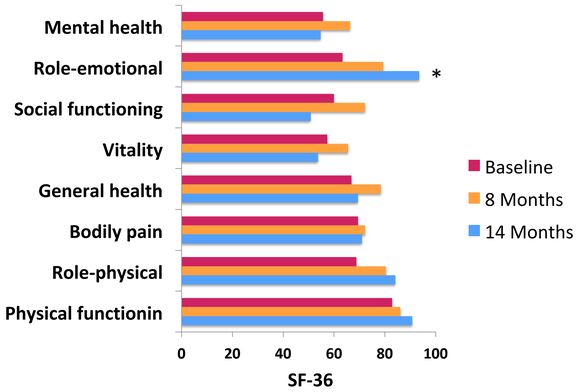

Figure 2. Quality of life assessed by SF-36 showed significant improvement only for the role emotional dimension, with mean difference of 35.68 (95% CI 16.67;54.42) at 14 months as compared to baseline.

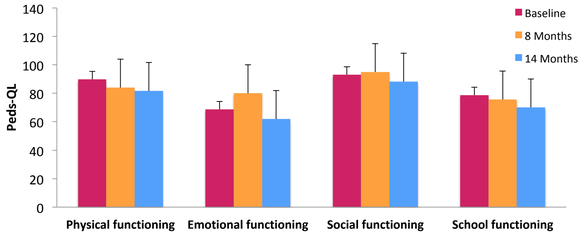

Figure 3. No significant changes in quality of life were observed among children (n = 8) using the Peds-QL questionnaire, within 8 and 14 months, as compared to baseline.

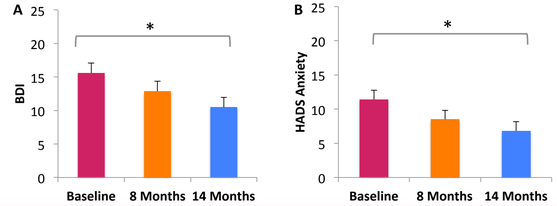

Figure 4. A. Significant decline in Depression assessed by BDI was seen at 14 months within intervention, as compared to baseline, with mean decrease of -6.17 (95% CI -12.28; -0.18). B. Improvement in Anxiety, assessed by HADS, with mean decrease of -4.38 (95% CI -7,04; -1,57) at 14 months within intervention, as compared to baseline, was also observed.

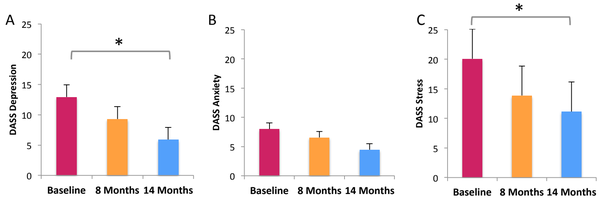

Figure 5. Significant decline in Depression and Stress assessed by DASS questionnaire was observed at 14 months within the study as compared to baseline, with mean decreases of -6.5 (95% CI -11.99; -1,14) and -8.87 (95% CI -13,78; -3.85), respectively.

Conclusion

Our results showed that a multidisciplinary approach to patients with HAE, addressing psychosocial and mental health in addition to medical aspects, resulted in improvement in quality of life, which is critical for best practice in HAE.

References

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