Objective assessment of nasality in Flemish adults with neurofibromatosis type 1

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Introduction

Hypernasality, a feature of velopharyngeal inadequacy (VPI), is often mentioned as characteristic of the speech of neurofibromatosis type 1 (NF1) patients. However, as Zhang et al. (2009) pointed out, the association between hypernasality, VPI, and NF1 has not been well elucidated. Studies into the nasality of NF1 patients generally relied on perceptual speech evaluations only, and those who did implement objective measurements either included patients suspected of VPI or lacked a control group. Therefore, the purpose of the present study was to document nasality in NF1 patients using an objective assessment technique (i.e., nasometry) and to compare their results with nasometric data obtained from a healthy control group.

Methods

Participants

The study group consisted of 30 Flemish NF1 adults, all fulfilling the diagnostic criteria for NF1, and 30 Flemish healthy controls frequency-matched for gender and age. As data was influenced by local dialect, only the participants living in East Flanders were selected for further analysis. Therefore, the final study group consisted of 24 NF1 adults and 16 controls (Table 1). None of the NF1 patients were diagnosed with pharyngeal, oral, or nasal tumors.

Nasometry

The Nasometer (Figure 1; Kay Elemetrics Corp., Lincoln Park, NJ, model 6200) was used to obtain nasalance scores. Participants were asked to sustain the speech sounds /a:/, /i:/, /u:/, and /m/ and read three reading passages designed specifically for use with the Nasometer. The first passage, an oronasal text, contains about the same percentage of nasal consonants as found in standard Dutch speech. The second passage, an oral text, lacks nasal consonants and the last passage, a nasal text, is by contrast overloaded with nasal consonants.

Results

NF1 patients versus controls

For all speech tasks, male NF1 patients displayed higher mean nasalance scores than male controls. However, values reached significance for the oronasal ($p = .035$) and oral text ($p = .022$) only. Mean nasalance scores of female NF1 patients were similar to those of female controls and differences were not significant at the level of $\alpha = .05$ (Figure 2).

Gender-related differences

Mean nasalance scores of male and female NF1 patients differed significantly from each other for the vowel /i:/ ($p = .006$) only.

Age-related differences

Male NF1 patients obtained higher mean nasalance scores with advancing age for the vowels /i:/ ($r = .715$, $r^2 = .51$, $p = .009$) and /u:/ ($r = .743$, $r^2 = .55$, $p = .006$).

Impact of MRI brain abnormalities

Based on the nasometric data, NF1 patients were split up into three subgroups: a normal, hyponasal, and hypernasal subgroup. Although the association between nasality subgroup and findings on MRI brain scan failed to reach significance ($p = .105$), it was interesting to see that all patients classified as hypernasal presented with MRI brain abnormalities (Table 2).

Conclusion

NF1 patients, males in particular, show a higher nasal resonance than healthy controls. Attention is attracted to the fact that all NF1 patients labeled as hypernasal presented with MRI brain abnormalities, suggesting a neurological basis and thus, problems with speech motor control. As such, subcortical structures, such as the thalamus and basal ganglia could be involved. A larger group of NF1 patients is needed to confirm this finding.

Table 1. Descriptive statistics of the final study group

	NF1 group (N = 24)	Control group (N = 16)
# Males/females	12/12	7/9
Mean (age) ^a	34.33	34.06
SD (age)	10.32	12.63
Min (age)	17	18
Max (age)	53	67

Note. Age is expressed in years.

^aAge distribution was not significantly different between male NF1 patients and male controls, nor between female NF1 patients and female controls.

Figure 1. The Nasometer

Figure 2. Boxplots of nasalance scores by reading passage, for males and females separately

Table 2. Distribution of NF1 patients by nasality subgroup and MRI findings

Subgroup	MRI brain abnormalities		Total
	Present	Absent	
Normal	4 (50%)	4 (50%)	8
Hypernasal	5 (100%)	0 (0%)	5
Hyponasal	2 (67%)	1 (33%)	3
Total	5	11	16

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