Voice and Swallowing Function in Neurofibromatosis 2

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INTRODUCTION

NF2 is a neuro-oncologic condition that presents with bilateral vestibular schwannomas (VS) of the cerebellopontine angle (CPA) and potentially schwannomas/ neurofibromas of other cranial nerves. Vagal injury can occur from direct involvement or as the result of surgical excision of CPA tumors. We assessed the prevalence of voice and swallowing impairments in this high-risk population. The present study aims to determine the rates and effects of vagal injury on voice and swallowing function in patients with Neurofibromatosis 2 (NF2).

METHODS AND MATERIALS

NF2 patients followed by a tertiary NF center were mailed Voice Handicap index and the Sydney Swallow Questionnaire surveys.

Evaluations:
1.Stroboscopic voice assessment
2.Swallowing evaluations by cine-fluoroscopy or fiberoptic endoscopy evaluation of swallowing (FEES)
3.Voice handicap index (VHI), which includes common voice complications and the negative effects on quality of life

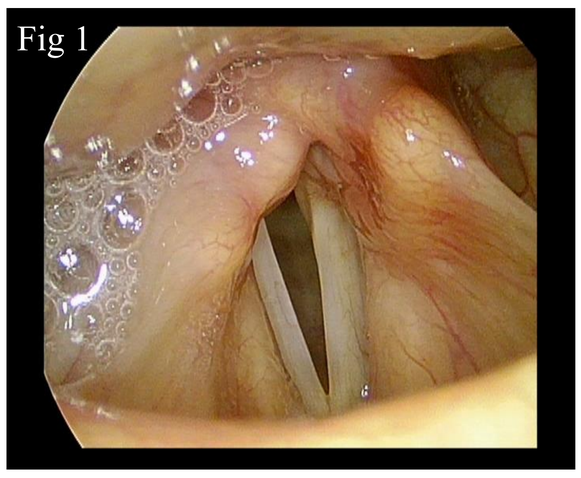

Figure 1: Vocal cord paralysis [right] with incomplete glottic closure. Note pooling of saliva in piriform sinus.

RESULTS

There were high rates of self-assessed and objective voice and swallowing handicaps in this population. 13/36 (36%) patients [VHI mean scores: abnormal - 66, normal - 7] patients have a self-assessed voice handicap and 20/35 (57%) patients have a self-assessed swallow handicap [SSQ mean scores: abnormal - 547, normal - 61]. Vocal cord paralysis/paresis (**Figure 1**) was observed in 15/18 (83.3%) patients examined with 20/36 (55.6%) of possible vocal cords affected. A strong correlation was found between vocal cord motion impairment and surgical intervention ipsilateral to the impairment (p=0.0022). 12/17 (71%) had abnormal GRBAS scores rated by blinded observers. Reflecting high vagal injury, there was reduced velopharyngeal closure (**Figure 2**) in 5/13 (39%), piriform sinus pooling (**Figure 3**) in 9/18 (50%), and abnormal swallowing functioning on FEES in 38% of patients. Pharyngeal residue is a risk factor for aspiration, and in one patient (12.5%) there was absent cough reflex with frank aspiration (**Figure 4**).

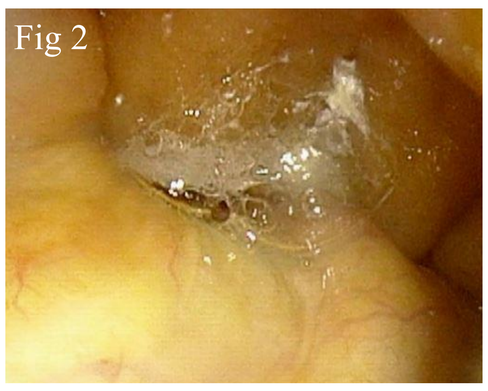

Figure 2: Velopharyngeal insufficiency. Bubbling of secretions in velopharynx secondary to incomplete palatal closure.

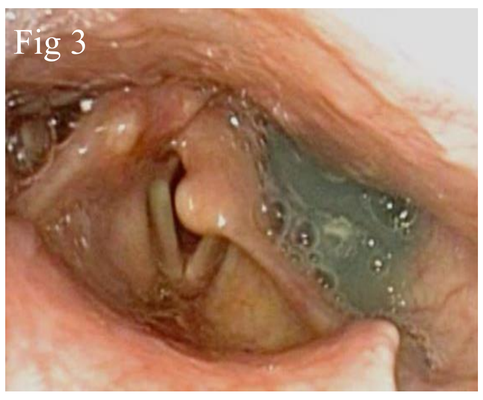

Figure 3: Fiberoptic endoscopy evaluation of swallowing (FEES), showing blue-dyed pharyngeal residue ipsilateral to vocal cord paralysis [left].

Figure 4: Cine-fluoroscopy [lateral view], demonstrating overflow aspiration in trachea (arrow).

DISCUSSION

Tumors of the CPA cause vagal dysfunction, which can lead to voice and swallowing problems and have a significant impact on a patient's quality of life. However, these issues are rarely reported. Previous quality of life studies have failed to mention voice and swallowing complications[1] even though there is a high prevalence of vagal palsy in patients with CPA tumors[2]. The consequences of vagal dysfunction, including speech and swallowing impairment, are evident in the present study. The findings in this study further support the importance of evaluating speech and swallowing function in NF2 patients in order to improve patient outcome and quality of life.

CONCLUSIONS

The results of this study demonstrate that speech and swallowing impairments are highly prevalent in NF2 patients and are most commonly related to surgical interventions in the CPA region.. The findings also point to the clinical importance of addressing these problems in order to provide patients with treatment options that can improve symptoms and prevent further medical complications.

REFERENCES

1) Neary WJ, et al. Use of a closed set questionnaire to measure primary and secondary effects of neurofibromatosis type 2. JLO. 2010 March 11;124:720-28

2) Best, SR, et al. Risk factors for vagal palsy following cerebellopontine angle surgery. Otolaryngol Head Neck Surg. 2012 August;147(2):364-368.