A Prospective Evaluation of Swallowing and Speech in Patients with Neurofibromatosis Type 2

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Sibi Rajendran[1] • Beth Solomon[2] • H. Jeffrey Kim, MD[3] • Tianxia Wu, PhD[4] • Gretchen Scott, RN[1] • Sarah Benzo, RN[1] • John Heiss, MD[1] • Prashant Chittiboina, MD[1]

[1]Surgical Neurology Branch, NINDS, National Institute of Neurological Disorders and S • [2]NIH Clinical Center Rehabilitation Medicine, National Institutes of Health Clinical Center • [3]NIDCD, National Institutes of Health Clinical Center • [4]Clinical Trials Unit, NINDS, National Institute of Neurological Disorders and S

Introduction: Neurofibromatosis Type 2 (NF2) is a multiple tumor syndrome of the central and peripheral nervous systems. In NF2, multiple schwannomas, ependymomas, and meningiomas cause neurological deficits from mass effect on adjacent neural structures, including hearing loss, tinnitus, balance problems, and cranial neuropathy. Deficits in speech and swallowing function are a significant source of morbidity in these patients, but these phenomena are poorly understood in NF2. Speech and swallowing deficits may arise due to the neuropathy of involved nerves, due to tumor growth, or as iatrogenic effects from neurosurgical procedures to remove these tumors. This prospective study aims to investigate swallowing and speech deficits in NF2 patients in the context of their natural history.

Methods: Imaging, clinical and speech/swallow data was prospectively collected on NF2 subjects through the Natural History of Neurofibromatosis 2 Study (NIH 08-N-0044). Patients were imaged with high resolution MRI pre/post Gadolinium contrast at each visit. Speech and oral motor function, along with cranial nerve exam was also performed by neurosurgery and neurotology physicians. The patients also completed a self-reported questionnaire that included responses to speech and swallowing functions. A Modified Barium Swallow (MBS) study (reported as ASHA Swallowing Independency Score from 1 - 7) was obtained from NF2 subjects who reported a speech or swallowing deficit on the questionnaire.

Results: Of the 168 patients enrolled in our study, 55 (33%, median age = 31, females = 38) reported subjective speech and/or swallowing deficits. These patients underwent one (n= 37) or multiple (n=18) MBS studies during 44.8 ± 10.4 month follow up. During MBS, a majority demonstrated near-normal swallowing (ASHA score >6, 82%), and no evidence of aspiration (aspiration/laryngeal penetration score =1, 96%). Prior to initial MBS consultation, 38 (69%) patients had undergone relevant neurosurgical procedures. In those with recent (< 1 week) posterior fossa surgery (n=12), 2 (17%) patients had severe dysphagia and high aspiration risk on post-operative MBS. Both of these patients recovered to functionally independent swallowing status (without any evidence of aspiration) within 12 days after surgery (one patient recovered within 24 hours).

Conclusions: Although, subjective complaints of swallowing or speech dysfunction are frequent in NF2, MBS demonstrates near-normal function, even in patients that have undergone multiple neurosurgical procedures. We suspect that this pattern reflects adaptive mechanisms in speech/swallowing in response to inexorable tumor growth and cranial neuropathy. Additionally, post-surgical swallowing deficits due to posterior fossa surgeries in NF2 patients are transient.