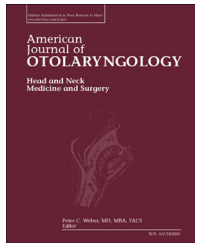
Available online at www.sciencedirect.com

ScienceDirect

www.elsevier.com/locate/amjoto



Is a redefinition of the growth criteria of vestibular schwannomas needed?

Dan Hougaard, MD[a,*], *Anne Norgaard, MD*[b], *Thue Pedersen, MD*[b], *Bo Martin Bibby, MSc, PhD*[c], *Therese Ovesen, DMSci*[a]

[a] *ENT Department, Aarhus University Hospital, Denmark*
[b] *Department of Neuroradiology, Aarhus University Hospital, Denmark*
[c] *Department of Biostatistics, Aarhus University, Denmark*

ARTICLE INFO

Article history:
Received 22 May 2013

ABSTRACT

Purpose: The natural history of vestibular schwannomas is poorly understood. Knowledge of growth rate and growth pattern is essential because the treatment strategy is based upon these. The purpose of this study was to determine the inter- and intraobserver variability in measuring VS size.

Materials and methods: Two consultant neuroradiologists independently made three linear measurements (d1, d2, d3) using digital MRI scans. MRI scans from 72 patients diagnosed between 2002 and 2010 with VS were obtained. These patients had a total of 223 MRI scans. d1 (medio-lateral diameter) was made perpendicular to d2. d2 was made parallel to the posterior border of the petrous ridge, and d3 was a measure of the cranio-caudal height of the tumor.

Results: Limits of Agreement ranges are larger for interobserver reliability compared to intraobserver reliability. Measurement error for all diameters (except d1, intraobserver) is greater than 2 mm. d1 measurements had the least variability and d3 measurements the highest variability, both for intra and interobserver measurements.

Conclusions: The optimal method of estimating VS size needs further investigation, and measurements need to be standardized and clearly defined. d3 seems to be the most difficult diameter to measure reliably. Interobserver measurement error for all diameters is greater than 2 mm. The current VS growth criterion of more than 1–2 mm, used to triage patients to surgery, lies within this error range, and thus is problematic as a guide for clinical practice. We therefore suggest that the growth criterion for VS be redefined.

1. Background

Unilateral vestibular schwannoma (VS) is a benign tumor arising from abnormally proliferative Schwann cells, which envelope the lateral portion of the vestibular nerve in the internal acoustic meatus [1]. The etiology of VS is poorly understood. Genetic defects, such as chromosomal 22q deletions [1], and infectious causes, such as herpes virus, have been implicated in the pathogenesis of VS [2].

With the exception of patients with type II neurofibromatosis, no genetic predisposition for VS has been identified. Neurofibromatosis patients, in contrast to those with sporadic Schwannomas, usually develop bilateral VS.

VS typically presents with asymmetrical, high-tone sensorineural hearing loss (75% of cases) together with a

* *Corresponding author*. ENT Department, Aarhus University Hospital, Building 10, 1st Floor, Norrebrogade 44, 8000 Aarhus, Denmark. Tel.: +45 61689062.
E-mail address: ddh@dadlnet.dk (D. Hougaard).

0196-0709/$ – see front matter © 2014 Elsevier Inc. All rights reserved.
http://dx.doi.org/10.1016/j.amjoto.2013.08.002

discriminatory loss. Sudden deafness is the initial symptom in about 10% of cases and the remaining 15% present with disequilibrium, unilateral tinnitus, cranial nerve symptoms or Ménière's disease-like symptoms [3].

The growth pattern of VS is multifaceted. VS may grow continuously or only to a certain size, followed by stagnation or even shrinkage [4]. Growth of VS leads to gradual filling of the internal acoustic meatus and, in some cases, protrusion out of the porus. Growth occurs within the initial 5 years following diagnosis; growth occurs in a limited number of tumors and primarily in those with an extra-meatal extension [4]. VS are for the most part slow growing tumors, but so far it has not been possible to predict the individual growth pattern. [1,5,6] Many studies have attempted to identify predictors of tumor growth, but have failed to identify any correlation between growth and variables such as age, laterality, sex and duration of symptoms [6]. One study found unilateral tinnitus to be predictive of tumor growth [7], while another has found tumor size at diagnosis to be predictive of tumor growth [8].

Knowledge of the VS growth rate and growth pattern is essential because the treatment strategy is based upon this [4]. In Denmark, VS surgery is centralized and performed at only one facility; Copenhagen University Hospital. Surgical indications include tumor size larger than 15 mm, tumor growth of more than 2 mm, and cystic tumors which may display sudden and dramatic growth [4]. If these criteria are not met, the patients are allocated to a "wait and scan" regimen in which the audiovestibular symptoms and VS growth are monitored regularly. VS growth is assessed by MRI (Fig. 1).

Reporting the size of VS is a challenge, which is reflected by the multitude of different classifications currently in use (at least 20 classifications are available) [1]. Gadolinium-enhanced MRI is the gold standard imaging investigation for tumors of the cerebellopontine angle, with a sensitivity of almost 100% and an extremely high specificity, capable of detecting tumors as small as 4 mm [9]. The traditional method of assessing tumor size is a single (axial) linear measurement assessing the greatest diameter. This method is also the one currently used in Denmark to register growth of VS [4,10,11]. However, several studies suggest that a more accurate way of describing VS growth is by volumetric measurement [12,13]. VS tumors grow in all directions and may be shaped in different ways, ranging from spherical to lollipop-shaped to dumbbell-formed. The shape may also change as the tumor expands into the intracranial space [13,14].

The incidence of VS in Denmark has been increasing during the last four decades. The annual number of diagnosed VS has increased from 15 cases in 1976 to 123 cases in 2004, corresponding to an increase in incidence from 3.1 VS per million per year to 22.8 VS per million per year in 2004. After 2005 a slight decrease in the number of diagnosed tumors has been noted, with 105 cases in 2008 (corresponding to an incidence of 19.4 VS per million per year) [10].

1.1. *Hypothesis*

A single (axial) linear measurement may not be the best way of assessing VS tumor size and growth. Growth occurs in three dimensions and the shape of the individual tumor is unpredictable [13–15]. Furthermore, other studies suggest that this way of measuring VS is subject to significant intra- and interobserver variability [15,16]. One study has shown an interobserver variability of 7.4 mm (Limits of Agreement, LoA) and an intraobserver variability of 2.5 mm when measuring the same tumor [16]. Yet, many authors consider a 1-2 mm increase in tumor size on a subsequent scan, based on two-dimensional

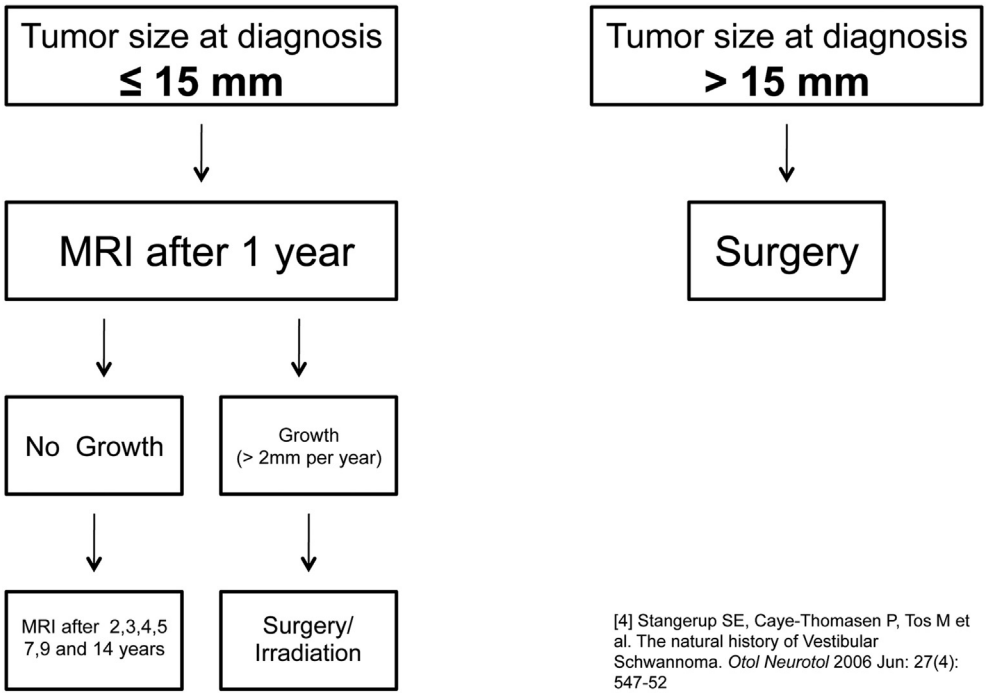


Fig. 1 – VS treatment strategy in Denmark.