Preview of the Medifocus Guidebook on: Acoustic Neuroma

Updated January 11, 2024



This document is only a <u>SHORT PREVIEW</u> of the **Medifocus Guidebook on Acoustic Neuroma**. It is intended primarily to give you a general overview of the **format and structure** of the Guidebook as well as select pages from each major Guidebook section listed in the Table of Contents.

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1 - Background Information

Introduction

Chronic or life-threatening illnesses can have a devastating impact on both the patient and the family. In today's new world of medicine, many consumers have come to realize that they are the ones who are primarily responsible for their own health care as well as for the health care of their loved ones.

When facing a chronic or life-threatening illness, you need to become an educated consumer in order to make an informed health care decision. Essentially that means finding out everything about the illness - the treatment options, the doctors, and the hospitals - so that you can become an educated health care consumer and make the tough decisions. In the past, consumers would go to a library and read everything available about a particular illness or medical condition. In today's world, many turn to the Internet for their medical information needs.

The first sites visited are usually the well known health "portals" or disease organizations and support groups which contain a general overview of the condition for the layperson. That's a good start but soon all of the basic information is exhausted and the need for more advanced information still exists. What are the latest "cutting-edge" treatment options? What are the results of the most up-to-date clinical trials? Who are the most notable experts? Where are the top-ranked medical institutions and hospitals?

The best source for authoritative medical information in the United States is the National Library of Medicine's medical database called PubMed®, that indexes citations and abstracts (brief summaries) of over 7 million articles from more than 3,800 medical journals published worldwide. PubMed® was developed for medical professionals and is the primary source utilized by health care providers for keeping up with the latest advances in clinical medicine.

A typical PubMed® search for a specific disease or condition, however, usually retrieves hundreds or even thousands of "hits" of journal article citations. That's an avalanche of information that needs to be evaluated and transformed into truly useful knowledge. What are the most relevant journal articles? Which ones apply to your specific situation? Which articles are considered to be the most authoritative - the ones your physician would rely on in making clinical decisions? This is where *Medifocus.com* provides an effective solution.

Medifocus.com has developed an extensive library of MediFocus Guidebooks covering a wide spectrum of chronic and life threatening diseases. Each MediFocus Guidebook is a

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high quality, up- to-date digest of "professional-level" medical information consisting of the most relevant citations and abstracts of journal articles published in authoritative, trustworthy medical journals. This information represents the latest advances known to modern medicine for the treatment and management of the condition, including published results from clinical trials. Each *Guidebook* also includes a valuable index of leading authors and medical institutions as well as a directory of disease organizations and support groups. *MediFocus Guidebooks* are reviewed, revised and updated every 4-months to ensure that you receive the latest and most up-to-date information about the specific condition.



About Your MediFocus Guidebook

Introduction

Your *MediFocus Guidebook* is a valuable resource that represents a comprehensive synthesis of the most up-to-date, advanced medical information published about the condition in well-respected, trustworthy medical journals. It is the same type of professional-level information used by physicians and other health-care professionals to keep abreast of the latest developments in biomedical research and clinical medicine. The *Guidebook* is intended for patients who have a need for more advanced, in-depth medical information than is generally available to consumers from a variety of other resources. The primary goal of a *MediFocus Guidebook* is to educate patients and their families about their treatment options so that they can make informed health-care decisions and become active participants in the medical decision making process.

The *Guidebook* production process involves a team of experienced medical research professionals with vast experience in researching the published medical literature. This team approach to the development and production of the *MediFocus Guidebooks* is designed to ensure the accuracy, completeness, and clinical relevance of the information. The *Guidebook* is intended to serve as a basis for a more meaningful discussion between patients and their health-care providers in a joint effort to seek the most appropriate course of treatment for the disease.

Guidebook Organization and Content

Section 1 - Background Information

This section provides detailed information about the organization and content of the *Guidebook* including tips and suggestions for conducting additional research about the condition.

Section 2 - The Intelligent Patient Overview

This section of your *MediFocus Guidebook* represents a detailed overview of the disease or condition specifically written from the patient's perspective. It is designed to satisfy the basic informational needs of consumers and their families who are confronted with the illness and are facing difficult choices. Important aspects which are addressed in "The Intelligent Patient" section include:

- The etiology or cause of the disease
- Signs and symptoms
- How the condition is diagnosed
- The current standard of care for the disease
- Treatment options



- New developments
- Important questions to ask your health care provider

Section 3 - Guide to the Medical Literature

This is a roadmap to important and up-to-date medical literature published about the condition from authoritative, trustworthy medical journals. This is the same information that is used by physicians and researchers to keep up with the latest developments and breakthroughs in clinical medicine and biomedical research. A broad spectrum of articles is included in each *MediFocus Guidebook* to provide information about standard treatments, treatment options, new clinical developments, and advances in research. To facilitate your review and analysis of this information, the articles are grouped by specific categories. A typical *MediFocus Guidebook* usually contains one or more of the following article groupings:

- *Review Articles:* Articles included in this category are broad in scope and are intended to provide the reader with a detailed overview of the condition including such important aspects as its cause, diagnosis, treatment, and new advances.
- *General Interest Articles:* These articles are broad in scope and contain supplementary information about the condition that may be of interest to select groups of patients.
- *Drug Therapy:* Articles that provide information about the effectiveness of specific drugs or other biological agents for the treatment of the condition.
- Surgical Therapy: Articles that provide information about specific surgical treatments for the condition.
- Clinical Trials: Articles in this category summarize studies which compare the safety and efficacy of a new, experimental treatment modality to currently available standard treatments for the condition. In many cases, clinical trials represent the latest advances in the field and may be considered as being on the "cutting edge" of medicine. Some of these experimental treatments may have already been incorporated into clinical practice.

The following information is provided for each of the articles referenced in this section of your *MediFocus Guidebook:*

- Article title
- Author Name(s)
- Institution where the study was done
- Journal reference (Volume, page numbers, year of publication)



• Link to Abstract (brief summary of the actual article)

Linking to Abstracts: Most of the medical journal articles referenced in this section of your MediFocus Guidebook include an abstract (brief summary of the actual article) that can be accessed online via the National Library of Medicine's PubMed® database. You can easily access the individual abstracts online via PubMed® from the "electronic" format of your MediFocus Guidebook by clicking on the corresponding URL address that is provided for each cited article. If you purchased a printed copy of a MediFocus Guidebook, you can still access the article abstracts online by entering the individual URL address for a particular article into your web browser.

Section 4 - Centers of Research

We've compiled a unique directory of doctors, researchers, medical centers, and research institutions with specialized research interest, and in many cases, clinical expertise in the management of the specific medical condition. The "Centers of Research" directory is a valuable resource for quickly identifying and locating leading medical authorities and medical institutions within the United States and other countries that are considered to be at the forefront in clinical research and treatment of the condition.

Inclusion of the names of specific doctors, researchers, hospitals, medical centers, or research institutions in this *Guidebook* does not imply endorsement by Medifocus.com, Inc. or any of its affiliates. Consumers are encouraged to conduct additional research to identify health-care professionals, hospitals, and medical institutions with expertise in providing specific medical advice, guidance, and treatment for this condition.

Section 5 - Tips on Finding and Choosing a Doctor

One of the most important decisions confronting patients who have been diagnosed with a serious medical condition is finding and choosing a qualified physician who will deliver high-level, quality medical care in accordance with curently accepted guidelines and standards of care. Finding the "best" doctor to manage your condition, however, can be a frustrating and time-consuming experience unless you know what you are looking for and how to go about finding it. This section of your Guidebook offers important tips for how to find physicians as well as suggestions for how to make informed choices about choosing a doctor who is right for you.

Section 6 - Directory of Organizations

This section of your *Guidebook* is a directory of select disease organizations and support groups that are in the business of helping patients and their families by providing access to information, resources, and services. Many of these organizations can answer your questions, enable you to network with other patients, and help you find a doctor in your geographical area who specializes in managing your condition.



2 - The Intelligent Patient Overview

ACOUSTIC NEUROMA

Introduction to Acoustic Neuroma

What is an Acoustic Neuroma?

Acoustic neuroma (AN), also called a vestibular schwannoma, is a benign tumor that is located on the eighth cranial nerve. The eighth cranial nerve, which traverses from the inner ear to the brain, is also called the auditory nerve or vestibulocochlear nerve. Auditory nerve cells, like many nerve cells in the brain and spinal cord, are wrapped or insulated by layers of specialized cells called Schwann cells. A gene on chromosome 22 directs Schwann cells to produce merlin, a protein that is also called schwannomin. Merlin acts as a tumor suppressor by preventing Schwann cells from rapidly dividing in an uncontrolled manner. When there is an abnormality or mutation of the gene, merlin is produced but is nonfunctional, and this enables the Schwann cells to rapidly multiply and form a tumor.

Acoustic neuromas are typically slow growing (approximately 2 mm to 4 mm per year) and usually develop over a period of years, although growth rates vary. These rounded tumors typically appear as a single mass and are encapsulated (confined to a contained area). At least 95% of acoustic neuromas are unilateral (occur on one side only). Unlike many other types of tumors, acoustic neuromas are benign and do not metastasize (spread) to other parts of the brain or the body.

Cranial Nerves and Acoustic Neuroma

Cranial nerves are a group of nerves that emerge directly from the brain. There are 12 cranial nerves, 10 of which emerge from the brainstem and 2 of which emerge from the cerebrum. The cranial nerves that are relevant to any discussion of acoustic neuroma are the auditory or vestibulocochlear nerve (eighth cranial nerve), the facial nerve (seventh cranial nerve), and in some cases, the trigeminal nerve (fifth cranial nerve).

The Auditory Nerve

The auditory nerve (eighth cranial nerve) is made up of two segments: the *vestibular nerve* segment and the *cochlear nerve* segment. The vestibular nerve transmits information about equilibrium (balance) from the inner ear to the brain. The cochlear nerve transmits information about sound. The vestibular nerve splits into two branches: the inferior (lower) and superior (higher) vestibular nerves. These two nerve components lie next to each other and next to the cochlear nerve as they pass through the *internal auditory canal*, a small bony canal that leads from the inner ear to the brainstem. They also lie next to the *facial nerve* (seventh cranial nerve). The



facial nerve activates the muscles that control facial movement. The vestibular portion of the auditory nerve is the site of origin in up to 95% of the cases of acoustic neuromas. Acoustic neuromas arise with equal frequency on the superior and inferior portion of the vestibular nerve.

The Internal Auditory Canal

The *internal auditory canal* is a small bony canal about 0.4 cm to 1.5 cm in length that is lined with the dura (the membrane that envelops the brain and spinal cord) and is filled with spinal fluid. It is the conduit for the eighth cranial nerve, the seventh cranial nerve, and blood vessels as they pass from the inner ear to the brainstem. As acoustic neuromas grow, they may protrude from the internal auditory canal (*extrameatal*) into the *cerebellopontine angle*, a spinal fluid-filled space that is located behind the temporal bone. Its boundaries include the brainstem, the cerebellum, and the temporal bone. This space provides the tumor with room to grow as much as 3 cm to 4 cm before it touches any important brain structures. An important blood vessel that passes through the cerebellopontine angle is the anterior inferior cerebellar artery (AICA), whose small branches provide the blood supply for the auditory nerve.

The Facial Nerve

The facial nerve (seventh cranial nerve) has motor and sensory functions. It controls the muscles that are responsible for facial expression, and also transmits taste sensation from the front two-thirds of the tongue and oral cavity to the brain. Facial nerve dysfunction in acoustic neuroma is usually a result of its being manipulated during tumor removal surgery.

The Trigeminal Nerve

The trigeminal nerve (fifth cranial nerve) is the largest cranial nerve and is responsible for sensation in the face, such as touch, position, pain, and temperature, as well as for motor functions, such as biting, chewing, and swallowing. The trigeminal nerve dysfunction is not common in acoustic neuroma but may occur due to pressure from the growth of the tumor or manipulation of the nerve during surgery.

Growth of Acoustic Neuromas

Acoustic neuromas typically originate at a point on the auditory nerve that is inside the internal auditory canal. The tumors may grow for many years before becoming symptomatic. They rarely shrink on their own and must, therefore, be monitored carefully after they have been diagnosed and then treated, if necessary. The growth rate ranges from no growth or very slow growth, to slow growth (0.2 cm per year on imaging scans), or rapid growth (greater than 1.0 cm/yr on imaging scans). Tumors may either consistently follow one growth pattern or may alternate growth rates at different periods of time. Tumors are usually described as "small" (less than 1.5 cm), "medium" (1.5 cm to 2.5 cm) or "large" (more than 2.5 cm).

Tumor growth rate one year following diagnosis is thought to be a strong predictor regarding the eventual need for treatment. Tumors that are larger than 2.5 cm at the time of diagnosis are considered more likely to grow than smaller size tumors. In addition, enlarged tumors may press on other nerves and alter facial sensation, or affect swallowing and vestibular functions, such as balance and hearing. Hearing loss, which occurs in approximately 90-95% of individuals with



acoustic neuroma, is usually caused either by compression of the cochlear nerve or of the blood vessels which supply the cochlea.

It is of utmost importance for individuals who are diagnosed with an acoustic neuroma to be under the care of a specialist. Health care professionals usually involved in the treatment of acoustic neuromas may include *otolaryngologists*, *neuro-otologists* (ear-nose-throat medical specialists with an additional subspecialty in conditions of the ear), *neurosurgeons*, *radiotherapists* (specialists in radiotherapy), and *audiologists*.

Incidence of Acoustic Neuromas

The incidence of acoustic neuromas is about one per 100,000 people. According to the National Institutes of Health (NIH), an estimated 2,000 to 3,000 new cases of acoustic neuromas are diagnosed in the United States each year. Reported incidence may be rising, however, due to improved imaging technology that enables smaller tumors to be clearly visualized. Because of the slow growth rate of acoustic neuromas, a large number of cases never become clinically evident, so that the actual number of cases may be much higher.

Acoustic neuromas account for approximately:

- 6% of all intracranial tumors (tumors occurring within the skull)
- 30% of all brainstem tumors
- 85% of tumors in the cerebellopontine angle region

Although they can occur as early as age 7, most people with acoustic neuromas are diagnosed between the ages of 30 and 60, with 50 being the median age at diagnosis. The incidence of acoustic neuroma is slightly higher among women (60%) than men (40%). Acoustic neuromas occur on one side of the head (unilateral) in approximately 95% of patients. The overwhelming majority of acoustic neuromas are due to unknown causes. Acoustic neuromas can also be associated with a genetic condition called *neurofibromatosis-2* (NF-2).

Neurofibromatosis-2 and Acoustic Neuromas

Neurofibromatosis-2 (NF-2) is a genetic condition that affects approximately 1 in 40,000 people (estimates range from 1 in 25,000 to 1 in 60,000) and is caused by a mutation in the NF 2 gene located on chromosome 22. Half of affected individuals inherit the disorder from an affected parent, and half appear to develop a spontaneous mutation of the gene with no identifiable cause. Symptoms of acoustic neuromas typically appear before the age of 21, in the teenage years and early adulthood. Bilateral presence of multiple acoustic neuromas is a prominent feature of NF-2. Approximately 2-4% of all acoustic neuromas are considered to be associated with NF-2.

Other features of neurofibromatosis-2 include:

- Meningiomas tumors that grow from the meninges (membrane covering the brain and spinal cord)
- Ependymomas tumors that line the passageways in the brain where cerebrospinal fluid is



produced and stored

• Ocular (eye) abnormalities - e.g., cataracts

Risk Factors for Acoustic Neuromas

Though the exact cause of acoustic neuromas is unknown, the only known genetic risk factor is one parent with neurofibromatosis type 2 (NF-2) that accounts for a minority of cases. Other conditions that have been identified that may place individuals at higher risk for developing an acoustic neuroma include:

- Prolonged noise exposure
- Mobile phones
- Childhood exposure to low-dose radiation of the head and neck
- History of a benign tumor of the parathyroid gland (in the neck)

While some studies have suggested that prolonged exposure to loud noise from sources such as power tools, construction machinery, or music, elevates the risk for developing acoustic neuromas, others have found that prolonged exposure to high noise levels does not raise the risk. Debate on this issue continues.

Speaking on mobile phones has received considerable attention as a possible cause of acoustic neuromas, since radiofrequency exposure from the phone is concentrated in the area closest to the handset, namely the ear and auditory nerve. Studies continue to be published regarding the possibility of an association between use of mobile phones and the development of acoustic neuromas. To date, an association has not been established and the debate continues. The most recent study was published in 2011 in *Cancer Epidemiology* in which the conclusion was that there was no increased risk of acoustic neuroma with regular use of a mobile phone or for users who began regular use 10 years or more before the study. More about this study can be viewed at: http://www.ncbi.nlm.nih.gov/pubmed/21862434

The Intelligent Patient Overview in the complete Medifocus Guidebook on Acoustic Neuroma also includes the following additional sections:

- Diagnosis of Acoustic Neuromas
- Treatment Options for Acoustic Neuromas
- Quality of Life and Acoustic Neuroma
- New Developments in Acoustic Neuromas
- Questions to Ask Your Health Care Provider About Acoustic Neuromas

To Order the Complete **Guidebook on Acoustic Neuroma** Click Here Or Call 800-965-3002 (USA) or 301-649-9300 (Outside USA)

3 - Guide to the Medical Literature

Introduction

This section of your *MediFocus Guidebook* is a comprehensive bibliography of important recent medical literature published about the condition from authoritative, trustworthy medical journals. This is the same information that is used by physicians and researchers to keep up with the latest advances in clinical medicine and biomedical research. A broad spectrum of articles is included in each *MediFocus Guidebook* to provide information about standard treatments, treatment options, new developments, and advances in research.

To facilitate your review and analysis of this information, the articles in this *MediFocus Guidebook* are grouped in the following categories:

- Review Articles 48 Articles
- General Interest Articles 29 Articles
- Surgical Therapy Articles 27 Articles
- Clinical Trials Articles 7 Articles
- Stereotactic Radiosurgery Articles 13 Articles

The following information is provided for each of the articles referenced in this section of your *MediFocus Guidebook:*

- Title of the article
- Name of the authors
- Institution where the study was done
- Journal reference (Volume, page numbers, year of publication)
- Link to Abstract (brief summary of the actual article)

Linking to Abstracts: Most of the medical journal articles referenced in this section of your *MediFocus Guidebook* include an abstract (brief summary of the actual article) that can be accessed online via the National Library of Medicine's PubMed® database. You can easily access the individual abstracts online via PubMed® from the "electronic" format of your *MediFocus Guidebook* by clicking on the URI that is provided for each cited article. If you purchased a printed copy of the *MediFocus Guidebook*, you can still access the abstracts online by entering the individual URI for a particular abstract into your computer's web browser.

4 - Centers of Research

This section of your *MediFocus Guidebook* is a unique directory of doctors, researchers, medical centers, and research institutions with specialized research interest, and in many cases, clinical expertise in the management of this specific medical condition. The *Centers of Research* directory is a valuable resource for quickly identifying and locating leading medical authorities and medical institutions within the United States and other countries that are considered to be at the forefront in clinical research and treatment of this disorder.

Use the *Centers of Research* directory to contact, consult, or network with leading experts in the field and to locate a hospital or medical center that can help you.

The following information is provided in the *Centers of Research* directory:

• Geographic Location

- United States: the information is divided by individual states listed in alphabetical order. Not all states may be included.
- Other Countries: information is presented for select countries worldwide listed in alphabetical order. Not all countries may be included.

Names of Authors

- Select names of individual authors (doctors, researchers, or other health-care professionals) with specialized research interest, and in many cases, clinical expertise in the management of this specific medical condition, who have recently published articles in leading medical journals about the condition.
- E-mail addresses for individual authors, if listed on their specific publications, is also provided.

• Institutional Affiliations

- Next to each individual author's name is their **institutional affiliation** (hospital, medical center, or research institution) where the study was conducted as listed in their publication(s).
- In many cases, information about the specific **department** within the medical institution where the individual author was located at the time the study was conducted is also provided.



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