

# Diagnosis of retrocochlear lesions with emphasis on expansion of the cerebellopontine angle

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The correct diagnosis of CPA tumours is a relatively common issue in both neurological and ENT practice, the omission of which can have serious consequences for the patient. Properly set clinical guidelines and diagnostic protocols are key aspects of good clinical practice. In the case of CPA tumours, two options are available: the first is diagnosis with the help of an ABR as the primary tool for determining the group of patients with a possible tumour; the second is an MRI scan of the posterior cranial fossa. With an appropriately set diagnostic protocol in place, and despite the 40% chance of failure of the ABR to detect tumours less than or equal to 1 cm, similar treatment results can be achieved with much higher cost efficacy in case of primary ABR testing.

**Key words:** CPA tumours, diagnosis

Received: November 13, 2017; Accepted with revision: March 22, 2018; Available online: April 17, 2018  
<https://doi.org/10.5507/bp.2018.013>

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## INTRODUCTION

Considering their most likely etiology, retrocochlear auditory lesions represent one of the prognostically more serious conditions and are potentially life-threatening. Clinical audiologists/otorhinolaryngologists play a key role by making an early diagnosis of these lesions. The most common lesion found in the posterior cranial fossa is vestibular schwannoma (VS), a benign tumour (occurring in one in every 5-7,000 patients), which accounts for approximately 70-80% of all tumours in the area. Its annual incidence is roughly 0.6-1.9 per 100,000 inhabitants. 95% of these tumours have a sporadic VS form, whereas 5% fall under the category of a neurofibroma (autosomal dominant hereditary form of type 2 neurofibromatosis (NF-2)). The mean age of patients with the sporadic VS form is 53 years, while NF-2 begins around the age of 25. In addition to VS, schwannomas of other nerves (most often n. V, less often n. VII and least often n. IX, X and XI), meningioma, cholesteatoma, lipoma and metastases are rarer findings in the cerebellopontine angle (CPA). Tympanojugular chemodectoma can spread to the cerebellopontine angle from the jugular foramen. Axial tumours of the posterior cranial fossa, such as ependymoma of the fourth brain chamber, medulloblastoma and many others, also penetrate the cerebellopontine angle<sup>1</sup>. Vestibular schwannomas are exclusively derived from the Schwann cells of n. VIII. During expansion of the tumour, the neuro-vascular bundle is compressed in the inner ear canal, mainly in the ventrocranial and ventrocaudal direction. This results in clinical symptoms of the disease. The growing tumour spreads through the path of least resistance

into the cerebellopontine angle where it has room for further growth; due to this further growth, the brainstem and its structures are then dislocated and compressed causing the symptoms associated to larger VSs. However, it can be completely inactive for many years and its growth rate varies from 0 to 20 mm per year. During the tumour's growth, we recognize 4 stages: intracanalicular, cisternal, brainstem compressive and hydrocephalic.

## DIAGNOSIS

Diagnosing tumours of the posterior cranial fossa and CPA is divided into 3 phases. The first phase aims to correctly identify patients suspected of having a tumour, thus beginning the diagnostic process in order to verify or exclude a tumour of the posterior cranial fossa. The second phase is the actual diagnosis based on a defined algorithm which is either based of audiovestibular findings<sup>2</sup> of direct MRI scanning. The third phase is a clinical audit and regressive evaluation of data, whether in terms of a possible late diagnosis or in terms of the effective use of resources in everyday clinical practice.

### Defining a group of patients suspected of having a CPA tumour

The most important factor in the diagnostic process for CPA and posterior cranial fossa tumours is determining the correct group of patients with a justified degree of suspicion of having a tumour, who will subsequently enter a more detailed diagnostic protocol. This group of patients needs to be broadly defined because they typical-

ly have a very vague clinical symptomatology, especially in the early stages of tumours in this area, but – due to the potential risk – the tumours must be diagnosed early enough so that the treatment still offers hope for a good quality of life and eventual low perioperative morbidity and mortality.

The most common symptom of a tumour of the posterior cranial fossa is hearing loss or tinnitus. The basic diagnostic tool used to screen patients suspected of having a CPA tumour is therefore a hearing test. A pure tone audiogram shows an asymmetrical hearing loss, which is considered the difference in hearing thresholds of more than 15 dB in at least 2 frequencies on the pure tone audiogram or a 15% difference in the hearing threshold for speech in the speech audiometry; however, CPA tumours should be excluded practically in any case of asymmetry in the area of hearing impairment including unilateral subclinical tinnitus without detected hearing impairment on the threshold audiogram. Asymmetric progredient hearing impairment occurs as the first symptom in about 80% of all CPA tumours; in about 8-10%, the first symptom is sudden hearing loss, which is usually explained by the interaction between the tumour and vascular supply. The third most common initial symptom is subclinical tinnitus without any detectable hearing impairment, which occurs in about 6-8% of the cases of CPA tumours (overall, tinnitus occurs in about 70% of all patients with CPA tumours) (ref.<sup>3</sup>). Only 2-4% of the patients with CPA tumours have a different presenting symptom, usually because that symptom is due to a pre-existing disorder that can cause hearing loss and that happens to co-exist with the problem resulting from compression of the nerve and its vascular supply due to a CPA tumour, e.g. a chronic middle ear inflammation. In this group of patients, the most common sign is dizziness, usually a mild form of dizziness with oscillopsia in the early stages of the tumour, which is usually induced by dynamic stress.

Therefore, the risk group of patients can be particularly characterized by:

- a) gradually deteriorating asymmetric sensorineural hearing loss
- b) sudden asymmetric hearing loss
- c) asymmetric tinnitus
- d) mild and otherwise inexplicable dizziness with oscillopsia, usually with a poor objective finding

## DIAGNOSTIC PROTOCOL

### Audiometric diagnosis

In the case of CPA tumours, the first diagnostic step is usually to do a topodiagnostic evaluation of the auditory function in terms of detecting retrocochlear hearing loss. However, retrocochlear hearing loss detection on its own, does not guarantee the presence of a tumour; the tumour is found in approximately 30-40% of the patients in whom a retrocochlear lesion is detected. Due to the large number of patients belonging to the group in which it is theoretically possible to assume the existence of CPA tumours, audiological topodiagnosis is the first step and a

basic diagnostic sieve that effectively eliminates patients in whom the CPA tumour is unlikely.

Theoretically, a physician can use the following methods to explain the reason for hearing impairment:

- a) tympanometry
- b) auditory evoked potentials

### A) Tympanometry

A tympanometric examination, unlike subjective audiometry, is objective; therefore, the validity of this examination is unquestionable. However, the value of a tympanometric examination for diagnosing a retrocochlear defect is limited. In the case of a retrocochlear disorder, we usually find a typical absence of the stapedial reflex due to a one-sided lesion in the centripetal arm of the reflex arc, when the ipsilateral reflex on the lesion side and the contralateral reflex on the opposite side are impaired. In the case of a subclinical lesion in the area of n. facialis, in addition to the ipsilateral reflex, we often find a deficit of the contralateral reflex on the affected side. With the cochlear type of sensorineural hearing loss, stapedial reflexes are usually present, even in cases of relatively considerable hearing loss due to the recruitment phenomenon. A more accurate variant of stapedial reflex testing is to examine the stapedial reflex decay; however, due to its considerable sensitivity to examination conditions, this method has not found a wide application in practice.

The importance of tympanometry lies primarily in the fact that it is a cheap and available method routinely used by most ENT doctors in the clinic. Tympanometry increases the degree of specificity with which we detect asymmetric sensorineural hearing loss; for example, it may point out the presence of conductive auditory disorder, which may sometimes be mistakenly diagnosed as a sensorineural disorder. Absence of a stapedial reflex can also be the first sign under which a physician suspects pathology of the inner ear canal or cerebellopontine angle. Tympanometry can therefore be perceived rather as a tool for screening patients entering the diagnostic protocol for CPA tumours more than a tool for detecting retrocochlear hearing impairment.

### B) Auditory evoked potentials

Introduction, sensitivity of the method

Hearing evoked potentials are the most accurate audiological method used to detect the presence of retrocochlear hearing loss. Currently, only brainstem auditory evoked potentials (ABR) are clinically useful. ABR was considered to be the most sensitive non-invasive method for detecting CPA tumours before the magnetic resonance age, with a sensitivity of about 98%. At the turn of the millennium, however, several published studies questioned the sensitivity of this method, especially in a subgroup of tumours smaller than 1 cm where ABR sensitivity is set at 58-82%. Therefore, the ABR method is being gradually dropped in some countries; as the preferred “gold standard”, an MRI of the brain is used instead as a screening method for detecting CPA tumours in high risk patients.

ABR sensitivity increases with tumour size. The method achieves 100% sensitivity in tumours larger than 2 cm and 89-92% sensitivity in medium-sized tumours between 1 and 2 cm. Using meta-analytical studies, the overall ABR sensitivity for detecting CPA tumours was determined as 93.4% (ref.<sup>4</sup>).

#### ABR limitations of use

The first limit of ABR use is the hearing threshold itself. Usually, ABR records show saturation (the phenomenon where the latencies of the wave response do not significantly decrease further with increasing stimulus intensity) when using a stimulus above 60 dB HL. Under typical settings, topodiagnosis is mostly performed taking two measurements on each ear at 70 and 80 dB HL. If the hearing thresholds in the tested frequency spectrum (i.e. between 2 and 5 kHz) occur at a level of intensity comparable to the stimulus intensity, then we cannot ascertain whether the absence of a response is due to the absence of the stimulation at the cochlea level in the intact conduction through the auditory nerve, or whether it is a primary disorder due to nerve suppression by the tumour. This is because in both cases the measurements can result in a completely desynchronized record. In such a case, when we cannot use a stimulus at least 20-30 dB higher than the auditory threshold, the ABR record is diagnostically unusable and the respective affected patient should always be recommended for MRI immediately. It might be

helpful to have a pathological finding on the contralateral record; however, this does not change anything in respect of the MRI indication.

The second case, when the ABR yield is reduced, relates to patients with hearing loss predominantly at higher frequencies. In such patients, the ABR response is delayed and desynchronizations occur not because of this phenomenon at the level of the auditory nerve, but because of the fact that the responses are predominated by the signal from apical parts of the cochlea where the stimulation – due to the propagation of the progressive wave – takes place later.

In order to compensate for this condition, a CE Chirp stimulus can be used instead of the broadband click; this will provide higher stimulation synchronicity even in the apical parts of the cochlea, thus increasing ABR sensitivity for small CPA tumours and tumours in the internal auditory canal<sup>5</sup>.

#### Examination by imaging methods

The gold standard for diagnosing CPA tumours is an examination using magnetic resonance imaging. MRI is the most sensitive diagnostic method, being capable of detecting tumours sized 2 mm or greater. Its main disadvantage is its high cost and, in some cases, its lower availability and the contraindications associated with it, the most important of which is an implanted pacemaker, cochlear implant or the presence of some metallic im-

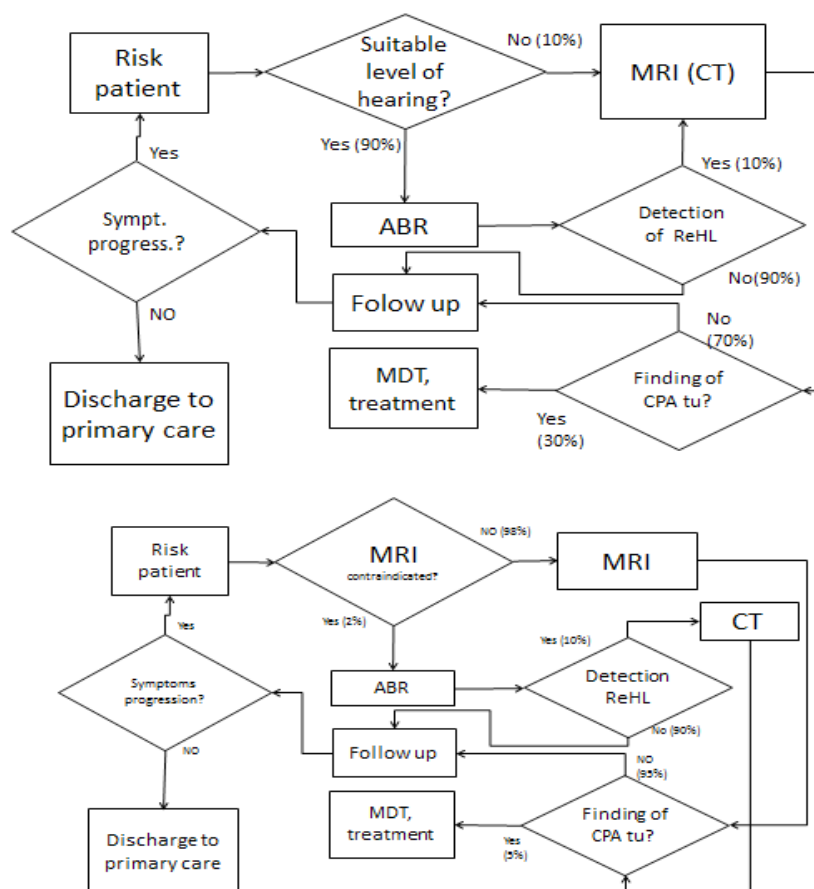


Fig. 1. Two forms of diagnostic algorithm for CPA lesions.

plants. When contrast agents are required, consideration should also be given to contraindications associated with impaired renal function or allergies to contrast agents.

The T2 weighted high-resolution image (T2W) or CISS (T2\*W) appear to be the most appropriate examination protocols. A CPA lesion is seen as a defect in the cerebrospinal fluid present in the cerebellopontine cistern. When a CPA tumour is suspected, T1W imaging together with the application of gadolinium contrast agent (GdT1W) is also recommended.

Using MRI imaging, it is also possible to distinguish the individual histological types of CPA expansion, especially meningioma, which has a more homogeneous structure and does not contain any cystic or haemorrhagic areas compared to schwannoma, which has a spongy structure and is usually accompanied by reactive saturation of the dura and is not accompanied by the extension of the internal auditory canal. An epidermoid cyst in the cerebellopontine cistern has a similar signal like cerebrospinal fluid, but with a marked diffusion restriction and a slight, non-homogeneous signal increase compared to cerebrospinal fluid in the FLAIR sequence (an image of soapy water); unlike real tumours, it does not saturate after the contrast agent is applied.

#### Retrospective analysis of patients entering the diagnostic protocol

As given in the foregoing previous text a clinician has two major choices for dealing with a patient whose clinical findings lead to suspicion of a CPA tumour. The two choices are shown in Fig. 1. If primary audiological diagnosis instead of MRI is selected and the selection of patients entering the diagnostic protocol is done correctly, an ABR examination should confirm the intactness of the auditory pathway in approximately 90% of the patients. For the remaining 10% of the patients, we should obtain an abnormal record, thereby necessitating an MRI examination. This MRI examination should subsequently confirm a tumour in the cerebellopontine angle in approximately 30% of the examined patients, which corresponds to about 3% of the incidence of CPA expansion in the entire group of patients entering the diagnostic protocol. A retrospective analysis should also specifically evaluate the group of patients with late-diagnosed tumours, i.e. tumours with a size greater than 2 cm, when the full spectrum of therapeutic options is no longer available and the therapeutic results are more often burdened by undesirable treatment complications and morbidity. In addition to determining the sensitivity of the diagnostic protocol, the result of these clinical audits should also include an analysis of the causes of the late diagnosis of CPA tumours.

## DISCUSSION

When designing clinical guidelines for diagnosing lesions in the cerebellopontine angle, two contradictory aspects have to be taken into account. The first aspect is the sufficient robustness of the diagnostic protocol from

the initial contact with the patient to the final diagnosis and treatment, so as to avoid a late diagnosis of tumours as much as possible. The second aspect is the emphasis placed on the price and availability of the examination methods.

At the end of the 1990s, ABR was an absolutely essential method for diagnosing retrocochlear hearing loss; this was undoubtedly related to the fact that MRI, the most sensitive method, was expensive and less accessible. However, studies and meta-analyses published early in this millennium<sup>6-9</sup> have reversed this, mainly noting the fact that ABR is not sensitive enough to diagnose small tumours sized below 1 cm, where there is almost a 40% probability of diagnostic failure. Nevertheless, even in countries with advanced health care systems and with a strong emphasis on preventing late diagnoses due to medical-legal reasons, e.g. the US, one-third of doctors dealing with the diagnosis of CPA tumours still use ABR as the primary screening test. Among other factors, this trend maybe attributed to the practice of private health insurance companies, which place great emphasis on the costs of examinations and thus regulate the use of financial resources.

In Western European countries with approved clinical guidelines, e.g. the UK with its NICE guidelines<sup>10</sup>, the ABR method is still listed as a relevant procedure for diagnosing CPA lesions; however, it is used only occasionally as a primary diagnostic tool, practically only when MRI is contraindicated (e.g. due to an implanted pacemaker) or if the patients themselves accept a lower yield of examination (e.g. because of claustrophobia). When significant asymmetric sensorineural hearing loss is detected, the patients routinely undergo MRI examinations under the basic T2W and T2\*W protocols without any contrast agent being applied. British authors<sup>11</sup> report that the specificity of MRI examinations under T2W and T2\*W protocols is comparable to the complete MRI examination protocol including T1W imaging with the application of gadolinium contrast agent, i.e. between 90-100%, and much cheaper. An important aspect affecting the choice of diagnostic procedure is undoubtedly the cost of the examination. In the US, the cost of MRI is significantly higher than that of ABR. In contrast in Western European countries, the cost and availability of these examinations are comparable (Danish authors<sup>12</sup> report EUR 319 for MRI and EUR 326 for ABR). This is one of the reasons why MRI is used as the primary examination in the overwhelming majority of cases in Western Europe; the ABR method is only of additional importance when MRI is contraindicated because there is no financial dilemma regarding the choice between diagnostic methods since the MRI is not more expensive, it is diagnostically superior and is as readily available as an ABR service.

In the Czech Republic, both approaches are used; MRI is generally preferred in high risk patients in large facilities where the MRI examination is readily available. If the facility is able to guarantee patients an acceptable waiting time for the examination, and the ABR examination method that focuses on the diagnosis of retrocochlear hearing loss is reserved only for cases where MRI



**Table 1.** A comparative pricing table for ABR and MRI in some countries.

| Country        | ABR                       | MRI                               |
|----------------|---------------------------|-----------------------------------|
| USA            | 230–540 USD (208–489 EUR) | 1,500–2,500 USD (1,360–2,270 EUR) |
| Denmark (EU)   | 326 EUR                   | 319 EUR                           |
| Czech Republic | 700 CZK (26 EUR)          | 7,000–14,000 CZK (260–520 EUR)    |

Recalculations are based on annual exchange rates of 2016 published by Ministry of Finance, Czech Republic  
(1 EUR = 27.04 CZK, 1 USD = 24.53 CZK)

is contraindicated. From this point of view, audiological methods excluding the pure tone threshold audiogram have ceased to play a primary role in diagnosing CPA tumours. Threshold audiometry of course as was already discussed, is necessary in order for the patient to enter the diagnostic protocol in the first place.

However if the efficient use of financial resources is taken into consideration, the choice of procedure is not straightforward. An MRI examination costs about CZK 7,000 on average, while an ABR examination is paid for by health insurance companies and costs CZK 700. For a complete MRI examination with all protocols, including the administration of the gadolinium contrast agent, the cost of the examination increases to CZK 14,000 per patient. Therefore, an MRI examination is approximately 10–20 times more expensive than ABR. Based on statistical data we should expect to diagnose 30 CPA tumours per 1,000 patients examined. According to published studies (situation), approximately 1/3 of these 30 patients will have a tumour sized less than 1 cm, where there is a 40% probability that ABR will fail to diagnose it; in patients with tumours between 1 and 2 cm, this probability is about 10%.

For every 1,000 patients with audiovestibular symptoms entering a diagnostic protocol using ABR as the primary screening method, there will be about 6 patients with false negative results. 2/3 of these patients will present with minor symptomatology and a tumour smaller than 1 cm, for which the subsequent indicated management – should be mostly clinical monitoring and follow-up with annual MRI scans. Therapy is generally started when clinical signs or MRI findings progress. Therefore, if a CPA lesion is missed in some patients and there is a progression of their clinical symptomatology, they are bound to eventually be re-examined audiotologically because of that and treated. Therefore, there is no great risk involved since CPA expansions grow slowly (about 2–20 mm per year), and many CPA expansions remain clinically and radiographically stationary for many years. It has been found that 50% of all tumours practically do not grow for years and that 5% of all tumours even show regression in their size in the follow-up MRI examinations<sup>13</sup>. The primary predictor of further tumour behaviour is its size at the time of diagnosis. Larger tumours are more likely to continue to grow, while small tumours typically do not grow at all or grow only very slightly. Therefore, it is likely that most tumours smaller than 1 cm will ultimately be just monitored with an annual MRI scan, and that no

treatment will be necessary for the overwhelming majority of them<sup>11</sup>.

It is apparent that with sophisticated and correctly clinically applied diagnostic protocols primarily based on audiological examinations, similar final results can be obtained in terms of timely treatment with a considerable amount of public health insurance funds saved thanks to the fractional costs of audiological diagnosis and missed opportunities. Similar findings were made by American authors in their meta-analytical study from 2013 (ref.<sup>14</sup>) as well as by authors from the Mayo Clinic<sup>15</sup>.

As things stand today the accepted protocol is to initiate CPA tumour therapy only after certain tumour size and growth dynamics are reached<sup>16</sup>, whereas tumours with a size below 1 cm are mostly just monitored<sup>11</sup> with respect to their growth dynamics, probable biological essence and clinical symptoms. We should therefore question whether it is necessary to diagnose tumours smaller than 1 cm, whose further management consists only of active surveillance. Especially in the case of the Czech Republic where the cost of a MRI scan is ten to twenty times higher than that of an ABR, then the debate about the suitability of the ABR as a cheaper and diagnostically almost as good method is fully justified. When the ABR examination is the primary method used to detect retrocochlear hearing loss, 90% of the patients entering the diagnostic protocol are excluded from having a CPA tumour, and only 10% of the patients undergo MRI. If we consider 100 patients without contraindications for ABR or MRI examinations, with an average price of one MRI examination being CZK 7,000 and the average price of one ABR examination being CZK 700, then CZK 140,000 is spent to investigate 100 patients suspected of having a CPA tumour when using ABR as the primary detection method. The prices of ABR and MRI procedures for USA, EU example of country and Czech Republic is given in Table 1. If all 100 patients were to be examined with MRI, this cost would increase to CZK 700,000. Based on known facts about the incidence and symptomatology of CPA tumours and the sensitivity of individual methods, when ABR is used as the primary method instead of MRI, the probability of a false negative result is approximately 16.5%. In the end, this represents approximately 1 per 200 patients entering the diagnostic protocol and approximately 1 patient per 6 patients with symptomatic CPA tumours. In the overwhelming majority of cases, however, the undiagnosed tumour will have clinical and biological parameters that do not require any intervention. It must be stressed that

doctors using more cost-effective but less sensitive methods will need sufficient legal protection in the event of lawsuits. Such legal protection can only be ensured by approved clinical guidelines.

Despite what has been mentioned above it is crucial for clinicians to realize that the observation of clinical guidelines does not relieve physicians of their responsibility to assess each patient and his/her risks individually, and intervene in a timely manner if they suspect any adverse development of the current condition.

## CONCLUSION

Despite better MRI sensitivity, both primary MRI examinations and primary audiological diagnosis using ABR are still equivalent diagnostic procedures, but only provided that they are applied correctly. Under Czech conditions, diagnostic protocols based primarily on audiological methods can lead to significant savings in public health insurance funds while keeping acceptable standards of diagnostic sensitivity.

### Search strategy and selection criteria

The objective of our research strategy was to evaluate the practice guidelines and common clinical practice in developed countries with emphasis on their differences across European countries and northern America. Scientific articles together with some keynote publications of well recognized authors and institutions were searched and evaluated using the PubMed database and Google search. The used phrases for a search were as follows: “vestibular schwannoma”, “acoustic neuroma”, “diagnostic efficacy”, “ABR” and “MRI”. Only papers in English were reviewed.

**Author contributions:** all authors – literature search, writing the manuscript, critical reading and manuscript revision.

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