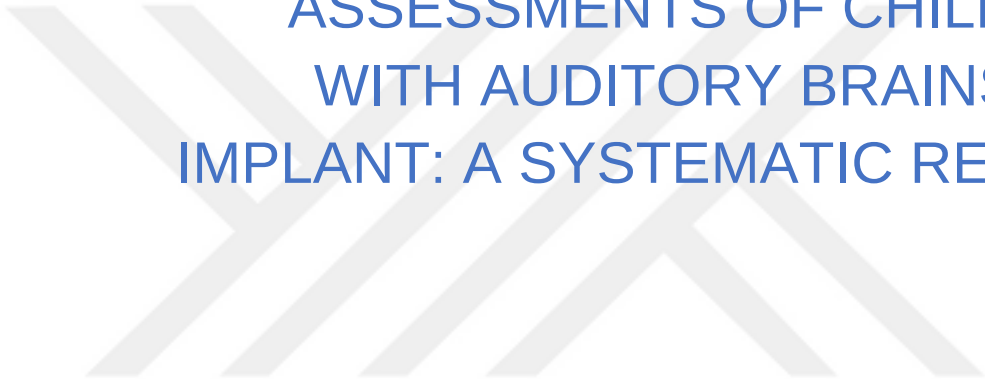




AUDITORY AND EDUCATIONAL ASSESSMENTS OF CHILDREN WITH AUDITORY BRAINSTEM IMPLANT: A SYSTEMATIC REVIEW



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It has been a long journey...

This dissertation dedicated to the memory of my grandmother, Samiye “Güngör” Teköz (1936-2015). You are gone but your belief in me has made this journey possible.

Abstract

This study, was conducted to determine the audiological, academic and other outcomes of Auditory Brainstem Implant (ABI) user children between 0-17 years old. In the study where systematic review method was used, 36 studies were included. The tools which were frequently used in these studies to assess audiological, academic and other developmental areas were determined and the outcomes were reviewed basically. Accordingly, it was determined that CAP, ITMAIS/MAIS, MUSS and Ling tests were the mostly used audiological assessment tools, that no assessment tools were used in the educational or other areas (n:31) but only in some (n:5) studies there are assessments and information related to these areas. Detailed statistical analyses of the outcomes could not be made due to non-homogeneous data. Consequently, it was seen that audiological outcomes were dominating in the majority of the studies but education area and other areas were ignored.

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1. Introduction

This study was conducted about how assessments in the audiological and educational areas are performed for 0-17 years old children with Auditory Brainstem Implant (ABI). As it is stated in the methodology section, the study was performed using systematic review method. Systematic review was basically executed to find answers to the following research questions and sub-questions:

- 1- How audiological assessments of the children with ABI are executed
 - a – Which assessment tools are used?
 - b – Which of these assessment tools are frequently used?
 - c – What are the basic outcomes obtained from the assessment tools used?
- 2- How educational or other developmental assessments of the children with ABI are executed
 - a --Which assessment tools are used?
 - b – At what frequencies these assessment tools are used in researches?

As it is seen, the research is focused on how the outcomes of medical implantations applied to individuals in a multidisciplinary area are obtained.

In the literature review section of this study, firstly a brief information about the history of ABI will be given to present information about its historical development. However, information about medical conditions and disorders to which ABI can be applied will be presented in Appendix because the content is too wide and technical. In that section, besides historical information, information about the studies obtained from previous studies will be presented. This information basically consist of the outcomes focused by the studies in the literature and the assessment tools they used. Also, information about the assessment tools used in the researches in the literature will be given. Thus, basic knowledge about ABI and children with ABI can be understood.

Finally, the systematic reviews in the literature will be mentioned and their results will be addressed basically.

In the methodology section, details about how this systematic review was conducted will be given. Firstly, we will mention what systematic review is and then explain why we preferred that method. Then, a review about how research restrictions (age group, disorders, anomalies etc.) have been determined will take place. In this section, there will be preliminary information about how the studies to be included in and excluded from the systematic review were determined. Thus, study design will be presented in detail.

In the following sections, the data analysis and outcomes, how the analyses have been made and the findings of analyses will take place. In the discussion section, the findings will be evaluated in detail and the answers of research questions will be discussed.

Within this systematic review, the concept of audiological outcomes will be grouped as all findings obtained from audiological and language development assessment tools whereas non-audiological findings are grouped as the outcomes of educational and other developmental areas.

2. Literature Review

First brain stem implant was applied in 1979 to a Neurofibromatosis type 2 (NF2) (for further information please see Appendices) female patient by William House et al. (House & Hitselberger, 2001). During the years following this application, brainstem implant could not be applied to non-tumor (not NF2 patient, for further information please see Appendices) pediatric patients due to various reasons. Among these reasons, the primary ones are the insufficiency of studies on the individuals (adults and children) who are not NF2 patients and the fact that the medical approval mechanisms in the countries like USA (FDA – Food and Drug Administration) have not approved pediatric applications until 2012.

First successful non-tumor pediatric ABI (Auditory Brainstem Implant) implantation was inserted by Colletti et al. to a child with auditory nerve aplasia in 2001. In the following years, the number of ABI implantations applied to non-tumor and NF2 patient children has increased rapidly. Just in a university in Turkey (Hacettepe University) more than 70 children without NF2 have been applied ABI until 2015 (Puram & Lee, 2015; Sennaroglu et al, 2009). One of the important reasons of the increase in the number of pediatric ABI studies after 2001 is the surgical success of pediatric ABI applications. However, surgically successful pediatric ABI implantations do not mean that ABIs will work efficiently in children. In other words, in order to understand how implants affect the audiological, communicative and academic skills of children, post-implant studies are needed. For example, pediatric cochlear implant (CI) (please see Appendices for differences cochlear implant and ABI) implementations have had a significant positive influence on the development of language, literacy, hearing skills and similar developmental areas of children (Spencer, Barker & Tomblin, 2003; Govaerts et al, 2002; Beadle et al, 2005; Geers, 2004). Systematic assessments of developmental areas are necessary in order to be able to prove above-mentioned developments in children.

The first outcomes of pediatric ABI implementations exhibit a wider variety than the first outcomes of CI implementations (Wilkinson et al, 2017). In some of the implementations dated early 2000s, the children were exhibiting essential audiological developments such as recognizing environmental sounds, recognizing

sound (Colletti et al, 2002) while in some studies children were exhibiting open-set speech recognition (Colletti, Shannon, Colletti, 2014). Another study involving 41 children with ABI implant between 2005-2013 revealed that all children acquired essential audiological perception skills whereas language and speech skills demonstrated a wide range of variety (Yucel, Aslan, Özkan & Sennaroglu, 2015). The reasons of this wide variety of ABI outcomes too demonstrate a variety. For example, child/adult's being or not being a NF2 patient (Colletti, 2006), type of surgical intervention approach for implantation (Wilkinson, Schwartz, Otto, & Brackmann, 2014; Puram, Herrmann & Barker, 2015; Colletti, Sacchetto, Giarbini, Fiorino & Carner, 2000), number of electrodes installed into brainstem (number of active electrodes) (Kuchta, Otto, Shannon, Hitselberger & Brackmann, 2004), position of electrodes installed into brainstem (Barber et al, 2017) and similar factors directly affect the outcome of pediatric ABI implementations. As it is understood, getting positive outcomes from a pediatric ABI implantation depends on simultaneous success of more than one parameter. Therefore, educational (academic), auditory and communicative skills of the children with ABI should be evaluated rigorously.

Looking at the researches and reports publishing the developmental data of children since the first implementation of ABI implantations, we see that the researchers have utilized specific tests and applications. Before mentioning the usage frequency of these test batteries in the literature, we should clarify a point. Particularly, majority of the studies reviewing the outcomes of pediatric ABI implementations have addressed the development of audiological and communicative skills whereas the studies about the affect of implant on the academic skills were nearly passed over.

Marangos et al. (2000), after the implementation of ABI implantation to a NF2 patient child, have performed the audiological evaluation of children directly through open-set tools. After the first pediatric non-tumor ABI implantations in 2001, the researchers have utilized fundamental phonetic and onomatopoeic sound detection for controlling post-implant development of children (Colletti, Fiorino, Sacchetto, Miorelli & Carner, 2001). The method used in this research intends to determine awareness of children with ABI towards environmental sounds and basic sounds. In the following researches, speech discrimination tools were added to audiometric test tools (Colletti et al, 2002). Thus, it was aimed to measure the degree of discriminating speech sounds by children after implantation.

In the research conducted during the following years, open-set and closed-set sentence and word recognition tests have been used (Sanna, Khrais, Guida & Falcioni, 2006; Colletti et al, 2004a). In the closed-set test batteries, children choose the words or sentences they hear among the choices within a certain set. On the other hand, in the open-set tests, the children try to determine the words or sentences they hear either from a large set of choices or without a set. The audiological and language test batteries used in the studies conducted after 2006 consist of tests with a more systematic structure. These are, CAP (Categories of Auditory Performance), ESP (Early Speech Perception), IT-MAIS (Infant Toddler Meaningful Auditory Integration of Sound), LIP (Listening Progress Profile), MAIS (Meaningful Auditory Integration of Sound) Ling 6 test, FAPCI (Functioning after Pediatric Cochlear Implantation), MSLDS (Manchester Spoken Language Development Scale), AuSpLan (Auditory Speech and Language), ELF (Early Listening Function), HINT (The Hearing in Noise Test), Identifying Early Phonological Needs in Children with Hearing Impairment, LIP (Listening Progress Profile), REEL (Receptive-Expressive Emergent Language Test), TELD-3 (Test of Early Language Development -3) evaluation batteries.

In the literature, CAP and MAIS/IT-MAIS are the tools which are mostly used in audiological assessment of non-tumor and NF2 pediatric ABI cases (Noij et al, 2015). CAP basically consists of 8 categories which involve auditory skills that get harder from 0 to 7:

0 = No awareness of environmental sounds

1 = Awareness of environmental sounds

2 = Response to speech sounds

3 = Identification of environmental sounds

4 = Discrimination of speech sounds

5 = Understanding common phrases without lip reading

6 = Understand conversation without lip reading

7 = Use of telephone with known speaker (Archbold, Lutman & Marshall, 1995)

MAIS/IT-MAIS is 10 questions, 4 categories survey by which parents evaluate the auditory behaviors of their children (Robbins, Renshaw & Berry, 1991). In addition to this, LIP is a profile covering awareness of environmental sounds to discrimination of speech sounds (Nikolopoulos, Wells & Archbold, 2000). ESP is a closed set discrimination test and picturing-pointing skill is necessary to implement the test (Eisenberg, Johnson & Martinez, 2005).

In the majority of studies conducted on children with ABI since 2000, audiological outcomes have been discussed. The most important reason of it is ABI implantation's being a medical intervention. The researchers dwell firstly on surgical and audiometrical outcomes following a medical process (ABI implantation).

Although with different reasons, information about the cognitive conditions of the children involved in the studies have been given and even some non-audiometric tests have been performed. Respectively Colleti and Zoccante (2008), Eisenberg et al (2008) and Sennaroglu et al (2009) have applied cognitive tests, intelligence tests and tests not measuring audiometric skills to the children involved in the studies. On the other hand, even in these studies, no information has been given about the conditions and academic skills of school-age children. In the studies other than that of Colleti ve Zoccante'nin (2008), non-audiometric tests and their outcomes have been used as contribution to audiometric assessments (Eisenberg et al, 2008; Sennaroglu et al, 2009). In the study of Colleti ve Zoccante'nin (2008), 17 children between 14 months and 16 years old have been applied LIPS-R (Leiter International Performance Scale-Revised). LIPS-R is a non-verbal cognitive test that can be applied when language differences prevent the measurement of cognitive skills of children whose language development is not satisfied. As it is seen, pediatric ABI studies have typically dealt with auditory and communicative outcomes but the studies dealing with cognitive development and the development of academic skills are not enough. Thus, it can be concluded that, in the literature, there are not enough studies assessing non-audiologic outcomes of the children with ABI.

Of course, there are studies that systematically review the outcomes of children with ABI (Noij et al, 2015; da Costa Monsanto, 2014; Merkus et al, 2014; Vesseur et al, 2018). In all of these studies, researchers have addressed the audiological outcomes of individuals with ABI (Noij et al, 2015; da Costa Monsanto, 2014; Merkus et al,

2014; Vesseur et al, 2018). Especially, in the studies of Noij et al. (2015) and da Costa Monsanto et al. (2014), audiological outcomes of the children with ABI have been directly handled. However, Noij et al. (2015) have excluded the outcomes of children who have hearing loss due to NF2 and dealt only with the outcomes of non-tumor children. Vesseur et al (2018) have compared the initiatives (CI or ABI) towards eliminating the hearing disability caused by Cochlear Nerve Deficiency using meta-analysis method. Merkus et al. (2014) have conducted a systematic review study comparing indications and contraindications of ABI.

However, these studies have been conducted in different areas than the areas of the answers of this systematic review's research questions in terms of both coverage and direction. First of all, in none of these studies, educational or non-audiological outcomes have been addressed. Furthermore, when we scan the literature, we can't come across any study mentioning that there are non-audiological outcomes which must be investigated separately. Due to these facts, above-mentioned studies (Noij et al, 2015; da Costa Monsanto, 2014; Merkus et al, 2014; Vesseur et al, 2018) have remained as just studies with medical focus. Naturally, there is a gap in this area.

In summary, since the installation of first ABI implantation, measuring the benefits of implants has been an important matter. Researches on the gains obtained by children with ABI implantation due to NF2 and non-tumor have accelerated in early 2000s. However, the studies have remained medically-focused and studies on the tests utilized in the researches are limited. However, it is possible to say that a wide variety of tests have been utilized in the studies. This systematic review aims at investigating the audiological tests applied in the studies in the literature and the educational and non-audiological gains referred by researches.

3. Methodology

In this study, systematic review methodology was used to find the answers of research questions. Before detailing the reason why systematic review methodology was used, we must talk about what systematic review is.

Systematic review is a synthesis study where all studies in the related area are comprehensively screened, several inclusion and exclusion criteria are used, a quality standard is set for the studies to be reviewed and the studies to be included are determined, usually for answering a question or solving a problem (Grove, Burns & Gray, 2014; Centre for Reviews and Dissemination [CRD], 2008; Higgins & Green, 2011; Karaçam, 2014). In addition, according to Shamseer et al. (2015), in order to minimize bias in systematic review studies, systematic methods are used. Accordingly, Shamseer et al. (2015) have defined the basic properties of systematic review studies as follows:

- An open and repeatable methodology and a clearly identified set of objectives
- A systematic research meeting eligibility criteria and attempting to determine all studies
- An assessment of the validity of the findings of studies included (such as the assessment of dependability on prejudice and cumulative estimates)
- Systematic presentation and synthesis of the properties and findings of the included studies.

As it is seen, systematic review must present open objectives, consider certain criteria in researching these objectives and certain standards should be set in order to be able to make a quality assessment to reduce bias in systematic review and to increase its validity and reliability. This systematic review can present more clear, reliable and less biased outcomes at the stage of synthesis.

Well then, why is it important for systematic reviews to present less biased, more open and reliable outcomes?

We can use an example to answer this question. Let's suppose that UCL (University College London) has successively become world's number one on the basis of subject between 2014-2018 at IOE's (Institute of Education) QS University Ranking system. Based on this information, we can assume that IOE's educational standards are high level and they deliver a high-quality education. On the other hand, it is possible to say that, this judgement/assumption is not evidence-based. Accordingly, coming to a judgement or conclusion based on a study of a single institute (in this case, QS University Ranking) limits the reliability of the conclusion and the usability of the information obtained (IOE's being world's number one between 2014-2018). Because, the information/research outcome/judgement delivered through a single channel is by no means reliable unless systematically reviewed and can be compared with multiple outcomes. That's why evidence-based studies are necessary in order to enhance the validity and reliability of the outcomes obtained from today's educational researches. Studies where evidence-based studies are systematically reviewed and organized are possible through systematic reviews and meta-analysis studies. Therefore, the studies in the education area should be made evidence-based just like those in the medical area. Also, educators need the use of scientific methods and evidence-based arguments when performing their studies. Hargreaves (1997) states it as: "Doctors and teachers are similar in that they make decisions involving complex judgements. Many doctors draw upon research about the effects of their practice to inform and improve their decisions; most teachers do not, and this is a difference. (p. 407)"

Consequently, the reason why systematic review methodology was used in this study is the fact that we are looking for systematic, open and evidence-based answers to the research questions. Another reason why systematic review was preferred in the study is to determine how much the outcomes of ABI implantation studies conducted on children in the medical area are effective in the education area and at the audiometric level.

This systematic review was made thanks to the implementation of a series of systematic strategies. These strategies were primarily designed in a way to allow searching for the answers of research questions. First of all, the target audience to be included in the systematic review and its characteristics were determined. This

determination was necessary to be able to establish the keywords and search strategy necessary to perform the research.

Thus, firstly the age limit of study population was determined: Individuals below 18 years old were assumed as children. This assumption was made based on the child definition in the UNICEF's "Convention on the Rights of the Child" dated 1989 (Unicef, 1989). In this way, the discrepancies in the definition of "child" in the researches from various countries were prevented by setting an age range. Afterwards, the definition related to the etiological origins of children who were applied implantation was clarified in order to be able to better present the outcomes. For the sake of better examining this issue, the following can be said:

-ABI is applied to persons to whom CI implantation can't be applied or cannot benefit even if it is applied (Schwartz & Wilkinson, 2017; Vincenti, Pasanisi, Guida, Di Trapani & Sanna, 2008; Shah, Kozin, Kaplan & Lee, 2016). Accordingly, the population of ABI users can typically be divided into two groups;

*NF2 Patients

*Non-tumor patients (not NF2 induced).

Other than the medical differences between these two groups, there weren't significant differences enough to exclude them from this systematic review. Therefore, when reviewing the outcomes in the light of research questions for ABIs implanted to children, considering the principle of processing as much data as possible, both groups were included in the systematic review. Besides, there isn't any reason that might require to exclude the NF2 patient children and non-NF2 (non-tumor) children. For all these reasons, all ABI implanted children below 18 were included in this review. Up to this point, the age range and characteristics of research population (all children implanted ABI due to NF2 and non-tumor) were described.

As the characteristics population that is the subject of the review is determined, the keywords required for the databases to be screened for studies for the systematic review have started to be established. The keywords were determined as "auditory brainstem implant", "auditory", "brainstem", "implants", "implant", "children", "pediatry", "auditory", "education", "cognitive", "outcomes". Then, we investigated in which databases these words can be screened for.

An issue was encountered at the stage of determining the databases to be screened. The ABIs which are subjected to the systematic review are technically medical devices and are implanted through surgical process. That is to say, ABI implantations pass through processes involving medical interventions and implementations. Naturally, the researches related to children with ABI and other studies are mostly placed into medical databases. As it is seen from the previous systematic review studies, similar studies have only conducted by screening medical databases (Noij et al, 2015; da Costa Monsanto et al, 2014; Merkus et al, 2012; Vesseur et al, 2018). However, using only medical databases as in the other studies (Noij et al, 2015; da Costa Monsanto et al., 2014; Merkus et al, 2012; Vesseur et al, 2018) could lead various problems. For example, depending only on the medical databases for the objective of evaluating educational and non-audiological outcomes which are among the objectives of this study, might have caused the outcomes to accumulate within a narrow area. Consequently, in order to overcome the medical database bottleneck, we utilized databases including educational researches which have not been used in the other studies.

The medical databases to be used were determined as MEDLINE, PubMed, Embase, Cochrane and the educational research databases were determined as EBSCO Education, ERIC. Additionally, Google Scholar search engine was used. Screenings were performed on the databases with the same keywords in English and Turkish. The searches were based on the date range of December 2000 to May 2018.

After determining databases, inclusion and exclusion criteria were determined in order to be able to decide whether or not to use the research found by screening in this systematic review. At this stage, following goals were determined to establish inclusion and exclusion criteria:

- To be able to gather the outcomes of ABI implanted children within an interval as large as possible.
- To be able to analyze in more detail the data to be obtained from studies.
- In order to collect data more efficiently, to be able to access full text of the researches about children with ABI.

- To be able to obtain clear information about children's age range and whether or not they are ABI-implanted.

In line with these purposes, **“all studies involving at least 1 child participant/patient with ABI (NF2 and non-tumor) were included in this systematic review.”**

All studies with following properties were identified as exclusion criteria and not included in this study:

- Those not giving information about the age of participants and about whether or not they are ABI implanted.

- Those studies for which full text could not be accessed or about which efficient information **(for example overlapping information, unclear or lack of information)** could not be obtained.

-All studies which could not be obtained from external sources or through databases.

By inclusion and exclusion criteria, it is determined how much the studies obtained through screening are suitable for this systematic review. With these criteria, it is intended to increase the number of studies included to make a broader review and on the other hand, the systematic review is systematically organized by means of certain criteria.

After determining inclusion and exclusion criteria, PRISMA flow diagram was used for final determination of the studies to be included in the systematic review and be analyzed (Moher, Liberati, Tetzlaff & Altman, 2009). This flow diagram essentially consists of four stages (Moher et al, 2009):

-Identification; the section where databases and the studies coming from other sources are specified. In this section, total number of studies takes place.

-Screening; in this section, all studies that came from sources are combined, repeating ones are removed. The studies reviewed and excluded are shown separately. (In this section, studies are not reviewed as full text).

-Eligibility; the studies reviewed in screening are assessed as full text. The articles which are reviewed as full text and not meeting inclusion criteria are removed with reason.

Included; the studies which fully meet the inclusion criteria and ready for review are shown.

The method of obtaining studies to be used for analysis in this systematic review are shown with PRISMA 2009 flow diagram (see Diagram 1).

After collecting the studies considered suitable for systematic review observing the inclusion and exclusion criteria, quality and evidence level of studies were determined. At this point, we should explain what study quality means and why it is important. According to Harrison, Reid, Quinn and Shenkin (2016), the quality of a study is divided into two as methodologic and reporting. That is to say, the quality of a study is about how and by which method it was conducted and how the content presents outcomes. As an example, if the findings of a study which generally has a good methodology, and which uses the method quite good are not clearly presented, the quality of that study might be questioned. In the systematic review studies, determining the quality of studies allows to see differences and similarities between the studies in the review. Therefore, using the study qualities in the systematic review ensures a more objective assessment. For this systematic review, quality assessment was established based on the quality criteria used in some previous systematic review studies. (Kuo et al, 2014; Noij et al, 2015).

1 Clear description of research design (eg, observational, prospective, or retrospective)
2 Well-defined description of consecutive samples
3 Well-formulated inclusion and exclusion criteria
4 Reporting of study withdrawal/lost to follow-up
5 Demographics of population described: age, sex, diagnosis, deafness, history, cochlear implant history (0, 1, or 2 items = 0 points; 3 or 4 items = 1 point; 5 or 6 items = 2 points)
6 Appropriate control or comparison group
7 Operative information mentioned: surgical technique, side of implantation and device (0 or 1 item = 0 points, 2 or 3 items = 1 point)
8 Follow-up rate and length described
9 Complications and/or Side effects addressed
10 Non-audiometric findings addressed
11 Audiometric findings addressed
12 Consistent audiometric tools utilized among study group

Table 1: Assessment Criteria for Quality of Studies

12 different criteria were determined for quality assessment (See Table 1). As Harrison et al. (2016) stated these criteria were established to separately score methodological and reporting properties. Except the 5th one, all of the 12 assessment criteria were given a point. The 5th criterion was separated into three groups within itself and these three groups were given points between 0 to 2. The studies whose criteria were given points were separated into three categories based on the total points they have collected. Those which collected 0-5 points were qualified as “low”, which collected 6-9 points were qualified as “moderate” and which collected 10-13 points as “high”.

After determining the qualities of studies to be included in the systematic review as low/ moderate/ high, we progressed to determine the level of evidence for studies. Before explaining how we carried out this stage, we should explain the meaning of the level of evidence and its importance. The level of evidence is a system ensuring hierarchical sorting of evidences involved within a (medical) study (Burns, Rohrich & Chung, 2011). Thanks to these levels, some deductions can be made about the reliability of study outcomes. As an example, because case studies involve limited number of participants and do not allow comparison with other studies, the evidences they present are at lower levels compared to systematic review studies. When the level of evidence is evaluated together with the quality of study more reliable and comprehensive information about the study can be obtained. In this study, the levels

of evidence of studies to be included in the review were organized based on Oxford Centre for Evidence-Based Medicine (OCEBM) 2011 Levels of Evidence study.

According to this chart, the levels of evidence of studies were divided into 5 categories (OCEBM, 2011). At the first level, there are systematic review studies and similar ones while at the last level (5) there are expert opinion/mechanism-based reasoning studies. Other studies are placed between the range of 1 to 5 based on the type of evidence they present and their content.

Here, we must state an important aspect of this study. No study was excluded because of quality or the level of evidence.

Consequently, the keywords “auditory brainstem implant”, “auditory”, “brainstem”, “implants”, “implant”, “children”, “pediatry”, “auditory”, “education”, “cognitive”, “outcomes” were screened over MEDLINE, PubMed, Embase, Cochrane, EBSCO Education, ERIC and Google Scholar databases for December 2000 – May 2018. The databases were screened both in Turkish and English and total 773 studies (see Diagram 1) were obtained. Also, 8 studies were obtained from other sources. Among these studies, the text which is originally French was included in the study (Couloigner et al, 2014). The full text of the study of Couloigner et al. (2014) was translated by a professional translator from original French text for future use. Total 781 articles (773 from databases and 8 through other sources) were reviewed. At this stage, the review was based on titles and abstract sections of the studies. At this screening stage, 715 articles were excluded from the systematic review and remaining 66 studies were prepared to review their full texts.

After reviewing the full texts, 30 studies were excluded due to;

- Unclear participants information (n = 23)

- Lack of outcome information (n = 3)

- Overlapping participants data (n = 4)

36 articles were prepared for analysis (see Table 2).

References	Design, Evidence Level, Quality	Participants n, Mean Age year month, (Range)	Follow-up	Audiometric tools used	Educational - Cognitive tools used	Surgical Approach	Indication	Additional disabilities
1 Marangos et al, 2000	RCR, 4, Moderate	n: 1; 16 (16)	9 - 80 months	Open-set discrimination of polysyllabic numbers	No	Translabrynthine	NF2 (n:1)	No
2 Colletti et al, 2001	RCR, 4, Moderate	n:2; 3.5 (3-4)	1-3 months	Fundamental phonetic and onomatopoeic sound detection	No	Retrosigmoid	Cochlear malformations (n:1), Cochlear nerve aplasia (n:1)	Unknown
3 Colletti et al, 2002	RCR, 4, Moderate	n:3; 3 (2-6)	1-12 months	Fundamental phonetic sound detection, detection / recognition of onomatopoeic sound, speech discrimination and onomatopoeic sound detection	No	Retrosigmoid	Cochlear malformations (n:2), Cochlear nerve aplasia (n:1)	Unknown
4 Kalamarides et al, 2001	CR, 4, Moderate	n: 1; 17 (17)	at least 12 months	Open disyllabic words identification test, Open set recognition test	No	Translabrynthine	NF2 (n:1)	No
5 Colletti et al, 2004b	RCR, 4, Moderate	n: 2; 5.5 (4.5-6.5)	40 days- 2 months	Detection / discrimination / identification of instrumental sound, discrimination / identification of onomatopoeic sounds	No	Retrosigmoid	Auditory neuropathy (n:1), cochlear nerve aplasia (n:1)	Unknown
6 Colletti et al, 2004a	RCR, 4, Moderate	n: 1; 16 (16)	1 month	Closed set word and open set sentence recognition	No	Retrosigmoid	Head trauma (n:1)	Blindness (n:1)
7 Otto et al, 2004	RCR, 4, Low	n: 11; 16.3 (12-17)	12-120 months	Sound identification, speech perception	No	Unknown	NF2 (n:11)	No
8 Colletti et al, 2005	RNCS, 4, Moderate	n: 8; 7.3 (1.2-17)	12 months	Closed set word recognition, open set sentence recognition	No	Retrosigmoid	Cochlear nerve aplasia (n:6) Auditory neuropathy (n:1), cochlear ossification (n:1)	No
9 Kamesweran et al, 2005	CR, 4, Low	n:1; 15 (15)	Unknown	Enviromental sounds, closed-set speech recognition	No	Retrosigmoid	NF2 (1)	No
10 Sanna et al, 2006	CR, 4, Moderate	n: 1; 12 (12)	8 months	Sound identification, Open-set bi-syllable words and sentences	No	Translabrynthine	Cochlear ossification (n:1)	Unknown
11 Colletti et al, 2008	RNCS, 4, Moderate	n: 24; 6.11 (1.2-16)	6-84 months	CAP, MAIS, MUSS, LIP	LIPS-R	Retrosigmoid	Cochlear nerve aplasia (n:18), Auditory neuropathy (n: 1), cochlear ossification (n:1), Cochlear malformation (n:4)	No
12 Eisenberg et al, 2008	CR, 4, Moderate	n: 1; 3.3 (3.3)	18 months	ITMAIS, ESP, Ling 6 test	KABC-II, Vineland-II, BASC-II	Unknown	Cochlear nerve aplasia (n:1)	No

13 Sennaroglu et al, 2009	RCR, 4, Moderate	n: 11; 3.6 (2.5-5)	1 - 15 months	MAIS / ITMAIS, MUSS, Ling 6 test, closed set word identification	Denver Developmental Screening Test II, . Stanford-Binet Intelligence Scale	Retrosigmoid	Cochlear malformations (n:11)	ADHD (n:2), Global developmental delay (n:1)
14 O'Driscoll et al, 2011	RCR, 4, Moderate	n: 6; 4.2 (3.2-5)	Unknown	Detection of environmental sounds and speech	No	Translabyrinthine	NF2 (n:6)	No
15 Choi et al, 2011	RCR, 4, Moderate	n: 7; 5.1 (1.5-9.3)	24 months	CAP	No	Suboccipital	Cochlear nerve deficiency (n:7)	Cognitive+visual (n:1), Cognitive only (n: 2)
16 Goffi-Gomez et al, 2012	RCR, 4, Moderate	n: 4; 4.2 (2.9-7.3)	12-30 Months	ITMAIS, ESP, MUSS	No	Retrolabyrinthine	Cochlear ossification (n:1), Cochlear malformations (n:3)	Unknown
17 Tan et al, 2012	CR, 4, Moderate	n: 1; 12 (12)	Unknown	Speech discrimination	No	Unknown	Cochlear ossification (n:1)	Unknown
18 Colletti et al, 2013	RCS, 3, Moderate	n: 21; 3.4 (1.7-10)	12 - 96 months	CAP	No	Retrosigmoid	Cochlear Nerve Deficiency (n:21)	Cognitive (n:2), Cognitive+motor (n:2), cognitive+visual (n: 1), cognitive+SLI (n:1), Down syndrome (n:1)
19 Bayazit et al, 2014	RCR, 4, Moderate	n: 12; 3.9 (2.2-6.3)	4 - 44 months	CAP	No	Retrosigmoid	Cochlear malformation (n: 9), cochlear nerve aplasia (n:1), cochlear nerve hypoplasia (n:1), cochlear ossification (n:1)	Global developmental delay (n:1), ADHD (n:2), Motor delay: (n:1),
20 Colletti et al, 2014	RCS, 3, High	n: 20; 1.4 (all < 3)	42-96 months	CAP, environmental sounds, open-set speech perception	No	Retrosigmoid	Cochlear nerve deficiency (n:20)	No
21 Siegbahn et al, 2014	RCR, 4, Moderate	n: 5; 6.8 (2-15)	3 weeks - 48 months	CAP	No	Translabyrinthine	NF2 (n:1), Cochlear nerve malformations: (n:3), cochlear ossification (n:1)	Unknown
22 Couloigner et al, 2014	CR,4, High	n:3; 8.8 (3 - 12)	6 - 12 months	HINT, CAP, MAIS, Open list word recognition	Just basic info.	Translabyrinthine	NF2 (n:2), Cochlear malformation (n:1)	No
23 Colletti, Shannon and Colletti al, 2014	RCR, 4, Moderate	n:64; 4.08 (9 mo - 16 years)	12 - 144 months	CAP	No	Retrosigmoid	NF2 (n:3) Cochlear malformation (n:8), Cochlear nerve aplasia+ (n:29), Cochlear nerve aplasia only (n:20), cochlear ossification (n:1), ossification (n:2), head trauma (n:1)	8 children had mild motor disability, 8 behavioral impairment, 16 cognitive (8 with cognitive+ other disabilities), 4 SLI and 2 visual impairment.
24 Puram et al, 2014	CR,4, Moderate	n:1; 1.4; (1.4)	4 months	ITMAIS	QOL	Retrosigmoid	Cochlear Hypoplasia (n:1)	No
25 Yucel et al, 2015	RCR, 4, Low	n:36; 6.3 for (3 - 14.5)	at least 12 months	FAPCI, CAP, SIR, TELD-3, MSLDS	No	Unknown	Cochlear aplasia, cochlear nerve aplasia	ADHD (n:5), Global development delay (n: 4), Pervasive development disorder (n:1)
26 Bayazit et al, 2016	RCR, 4, Moderate	n:1; 17 (17)	Unknown	Ling 6 test	No	Retrosigmoid	Cochlear ossification (bacterial meningitis) (n:1)	No

27 Lundin et al, 2016a	CR, 4, Moderate	n:4; 2.8 (2 - 4)	3 - 78 months	CAP, Ling 6 Test	No	Translabyrinthine (n:3), retrosigmoid (n:1)	Goldenhar syndrome (n:1), CHARGE Syndrome (n:1), Cochlear ossification (n:1), Cochlear aplasia (n:1)	Unknown
28 Lundin et al, 2016b	CS, 4, Moderate	n:2; 7 (6 - 8)	Unknown	Questionnaire (Perception of environmental sounds and the benefit of ABI)	No	Unknown	Cochlear ossification (n:2)	Unknown
29 Puram et al, 2016	CR, 4, High	n: 4; 20 months (11 - 30 months)	3- 12 months	ELF	No	Retrosigmoid	Cochlear nerve aplasia (n: 4)	No
30 Sennaroğlu et al, 2016	RCR, 4, Moderate	n:35; 1.7 (1 - 5.5)	at least 12 months	CAP, FAPCI, MSLDS, SIR, Closed set- Open Set tests	No	Retrosigmoid	Cochlear hypoplasia (n:11), Michel deformity (n: 5), Cochlear aplasia (n: 6), Common cavity: (n:7), incomplete partition type I (n: 3), CAS* (n:3)	No
31 Wilkinson et al, 2017	CR, 4, High	n:5; 3.3 (3 - 4.11)	36 months	ITMAIS/MAIS, ESP	No	Retrosigmoid	Cochlear nerve deficiency (n:7), cochlear hypoplasia (n:1), cochlear aplasia (n:1)	No
32 Choi et al, 2017	RCR, 4, Moderate	n:9; 5.1 (1.5 - 9)	4 - 56 months	CAP, ITMAIS, MUSS, Ling 6 Test, word identification and sentence recognition test, daily sentence in Korean	No	Suboccipital	Cochlear nerve deficiency (n:9)	Blindness + cognitive (n:2), cognitive (n:1)
33 Jeyaraman et al, 2017	RCR, 4, Moderate	n:5; 4.11 (1.11 - 7.10)	12 -36 months	CAP, SIR, REEL, AuSpLan	No	Retrosigmoid	Michel's aplasia (n: 4), Cochlear nerve deficiency (n: 1)	Unknown
34 Jung et al, 2017	RCR, 4, Moderate	n:10; 3.9 (1 - 9)	15 - 88 months	CAP, ITMAIS, MUSS	No	Retrosigmoid	CHARGE syndrome (n:3), Cochlear aplasia (n:2), other/unknown: (n:5)	Cognitive (n:3), blindness (n:1)
35 Teagle et al, 2018	PCS, 4, High	n:5; 3.38 (2.1 - 5.5)	30-36 months	ITMAIS/MAIS, ESP, Identifying Early Phonological Needs in Children with Hearing Impairment	No	Retrosigmoid	CHARGE Syndrome (n:2), Michel's aplasia (n: 1), Cochlear nerve deficiency (n:2)	No
36 Malerbi et al, 2018	RCR, 4, Moderate	n:1; 11 (11)	at least 12 months	FC SS (suprasegmental four-choice test), FC (four-choice spondee test), Vowel (vowel/consonant/vowel presentation)	No	Retrolabyrinthine	Cochlear ossification (bacterial meningitis) (n:1)	Unknown

Table 2: Included Studies and Basic Contents,

Abbreviations: CR: Case Review, , C: Case Study, RCS: Retrospective Case Study, RCR: Retrospective Case Review, PCS: Prospective case-control study, RNCS: Retrospective nonrandomized case-control study, ESP: Early Speech Perception; IT-MAIS: Infant Toddler Meaningful Auditory Integration of Sound, LIP: Listening Progress Profile, MAIS: Meaningful Auditory Integration of Sound, MUSS: Meaningful Use of Speech Scale, ELF: Early Listening Function, SIR: Speech Intelligibility Rating, REEL: Receptive-Expressive Emergent Language Test, TELD-3: Test of Early Language Development -3, CAP: Categories of Auditory Performance, FAPCI: Functioning after Pediatric Cochlear Implantation, MSLDS : Manchester Spoken Language Development Scale, ,AuSpLan: Auditory Speech and Language, HINT: The Hearing in Noise Test, NF2: Neurofibromatosis Type 2

*There is no data for CAS abbreviation in original article.

4. Data Analysis and Results

The demographic information and other information of the children involved in the systematic review were organized using descriptive statistics methods and tools (SPSS 24.0). The outcomes of the studies were not statistically analyzed since they didn't have a homogeneous structure. Again, they were analyzed using descriptive statistics methods and tools (SPSS 24.0).

The articles to be included in the systematic review were subjected to quality assessment based on their contents and given points between 0-13. Thus, their qualities were determined as low, moderate or high. In order to determine their level of evidence, the studies were classified again based on research design using Oxford CEBM classification. Thus, the studies could be categorized as in the example below (Table 3):

References	Design, Evidence Level, Quality
Choi et al, 2011	RCR, 4, Moderate

Table 3: Example of categorization

In this organization, study design (RCR), the level of evidence (4) and quality (moderate) of studies were written separately. Among 36 studies, 28 (%78) were determined as moderate, 5 (%14) were determined as high and 3 (%8) were determined as low quality. Besides, the levels of evidence of 34 (%94) studies were defined as 4th level and only of 2 (%6) studies were as 3rd level. Following this process, in order to determine the demographic status of the participating children, number of participants, their gender and age were determined. For some studies, the number of study participants and the number of participants whose outcomes could be obtained were different. For example, for one study, total number of participants was X but the number of participants whose outcomes could be obtained was X- 1 (participant's outcomes cannot be obtained/does not exist). In such situations, the numbers giving information about participant outcomes were assessed.

4.1. Demographic Information of the Participating Children

With the arrangements made, total number of individuals participated to the studies were determined as 328. Their basic information and other information about the studies were listed in Table k. The gender distribution of 328 children were identified as 106 (%32) female, 93 (%29) male and 129 (%39) undefined. The highest percentage of the participant gender was female.

The ages of all children who participated to systematic review were obtained. Thus, average age of 328 children was determined as 4,9. Their ages were distributed between 9 months and 17 years old.

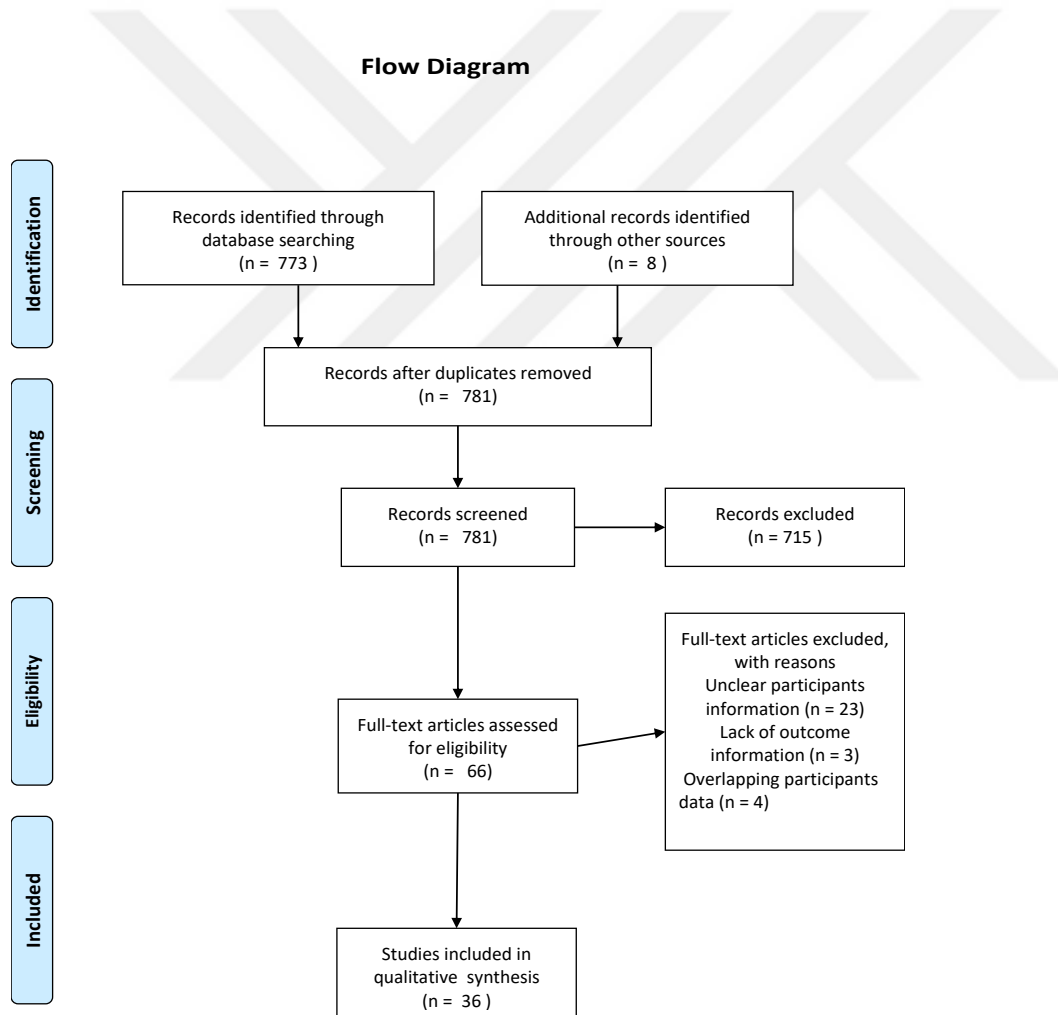
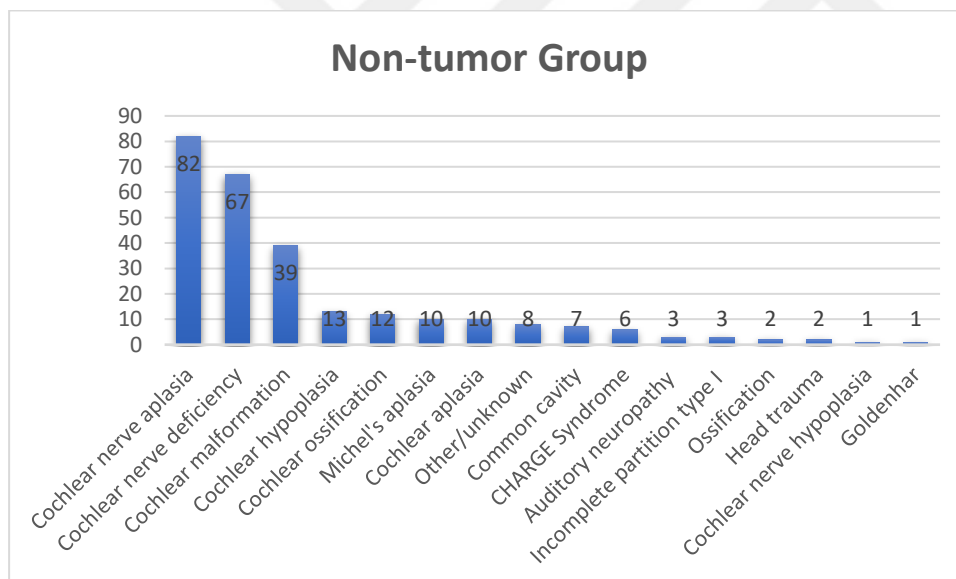


Diagram 1: Flow Diagram, Based on Moher et al, (2009)

4.2. Medical Information of the Participating Children

The children with ABI implantation have hearing loss basically because of two indications: NF2 and non-tumor anomalies and disorders. It was determined that, 26 (%8) children in the research have lost hearing due to NF2 and 302 children due to non-tumor, who needed ABI implantation.

On the other hand, we must emphasize that non-tumor group (n:302) does not have a homogeneous structure. In this group “Cochlear nerve aplasia” disorder is seen mostly (see Graphic 1). Only Yucel et al. (2015), have not indicated numerically the disorders and anomalies in this group. On the other hand, it is certainly known that each individual in this group of 36 has either Cochlear aplasia or cochlear nerve aplasia. Also, the indication of 8 children was specified as unknown/other. As it is seen, majority of this systematic review consists of children with non-tumor disorders.



Graphic 1: Children with non-tumor disorders

4.3. Additional Disabilities of Participating Children

In some studies, the participating children are reported to have additional disorder(s) besides NF2 and non-tumor disorders. Accordingly, in 16 studies the children are reported not having additional disabilities but in 9 studies the children have additional disabilities (other than hearing impairment). In 11 studies, no information is given. In 64 studies where children are reported to have additional disability, it is emphasized

that 64 children have one or more disabilities other than the hearing impairment. (16 of these children have more than one disability such as visual + motor disabilities). Here, before giving number of additional disabilities and related information, we should remind that, the classification and naming of disabilities are completely based on the information in the related study. Thus, some terms and naming do not match the terms and statements in today's literature (see Table 4).

Additional disabilities	n
Cognitive/intellectual disability (Mental Retardation)	23
ADHD	9
Blindness/Visual impairments	8
Emotional and Behavioral Disorders	8
Global development delay	6
SLI (Specific language impairment)	5
Motor disabilities	3
Down syndrome	1
Pervasive Developmental Disorders	1
Total	64

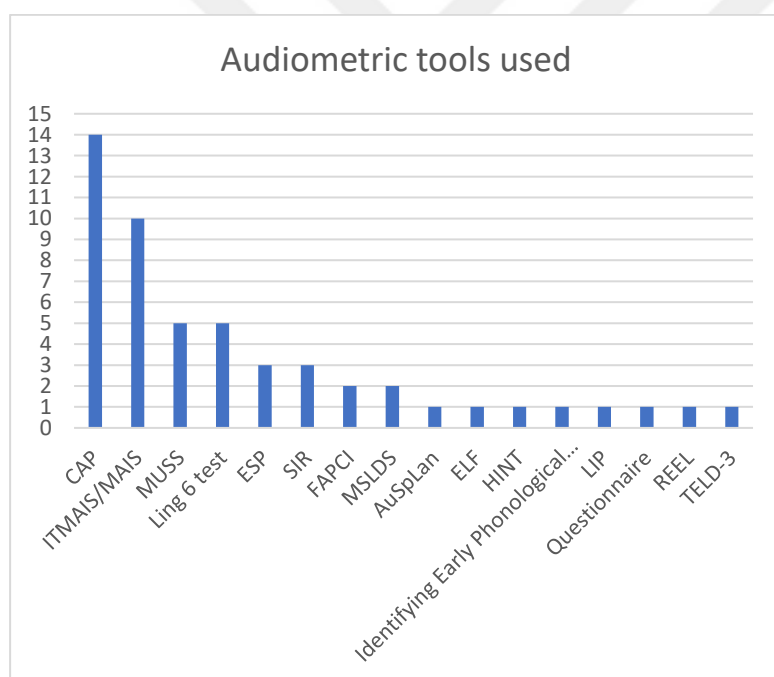
Table 4: Additional Disabilities of Participating Children

All of the participating children have had a surgical operation for ABI implantation. Since in the literature it is reported that some surgical intervention techniques give more effective results on NF2 patients (Colletti et al, 2000), the method used in ABI implantation is particularly indicated in most of the studies. Accordingly, in 21 (%57) studies retrosigmoid approach, in 7 (%19) studies translabyrinthine, in 2 (%5) studies retrolabyrinthine and in 2 (%5) studies suboccipital approaches were preferred. In 5 (%14) studies, no information was given about the surgery technique.

The follow-up periods during which children were followed-up post ABI implantation distribute over a large scale. In 31 studies, follow-up periods range between minimum 3 weeks and maximum 144 months. In 5 studies, no related information was found.

4.4. The audiometric Tools Used in and Audiometric Outcomes Obtained From Studies

The studies included in the systematic review have used a wide variety of tests when evaluation the audiological findings. It makes difficult to make data analysis over a non-homogeneous data set. Thus, when reviewing the audiological findings of the studies, primarily the tests used and their numerical distribution were reviewed. Accordingly, CAP was the mostly used (n:14) assessment tool. ITMAIS/MAIS was used in 10 studies and MUSS and Ling 6 Test batteries were used in 5 studies each. Other test tolls were used in different ratios (see Graphic 2). In this systematic review, average scores obtained from mostly used evaluation tools were found. The outcomes of other studies were not evaluated separately but general outcomes were handled.



Graphic 2: The audiometric Tools Used In Studies

In 10 studies, single test tool was used whereas in 26 studies multiple test tools were used (see Table 2). Here, it is important to emphasize the non-systematic test tools which are classified as “others” in this study. These are the tools that take place in the studies included and which are explained in detail by the researchers. The most important reason that they are classified as others is that these evaluation tools do

not have content enough to categorize them. These evaluation tools were used in 19 studies together with other systematic test and evaluation tools (see Table 2). In this systematic study, average scores obtained from mostly used evaluation tools were found. The outcomes obtained from other studies were not evaluated separately but general outcomes were handled.

In 11 of 14 studies where CAP evaluation was used (see Table 2) scores and average values could be found. Accordingly, in 4 of 11 studies (Sennaroğlu et al, 2016; Jeyaraman et al, 2017; Colletti et al, 2008; Colletti, Wilkinson, & Colletti, 2013) participant averages were found as 4 points or above while the scores varied between 1 and 3,3 in other studies. In the studies where average scores were not found, CAP scores are presented together with other values (such as ABI usage period) and just scores could not be obtained.

In 4 of studies where ITMAIS/MAIS evaluation was used, scores could not be obtained clearly whereas in the remaining 6 studies, the score averages were determined as 13,14/40. In the studies where ITMAIS and MAIS scores could not be evaluated clearly, it was seen that the results were presented as percentages rather than points and the results of MAIS and ITMAIS evaluations are mixed.

From 5 studies where MUSS evaluation was used (Colletti et al, 2008; Sennaroğlu et al, 2009; Goffi- Gomez et al, 2012; Jung et al, 2017; Choi et al, 2017) 2 of them do not give score information whereas the study of Colletti et al. (2008) give the score as percentage. Only in two studies the MUSS scores of the participants are given exactly (Sennaroğlu et al, Jung et al, 2017). In this study, only 2 users could take 30 or above points but the other (n:3) could not obtain 15 or above scores.

Ling 6 test was determined to be used in 5 different studies (Sennaroğlu et al, 2009; Jung et al, 2017; Eisenberg et al, 2008; Bayazit et al, 2016; Lundin et al, 2016a).

The evaluations for which number of use in the studies is ≤ 3 (ESP, SIR, FAPCI, MSLDS, AuSpLan, ELF, HINT, Identifying Early Phonological Needs in Children with Hearing Impairment, LIP, REEL, TELD-3) were not used in the outcome analysis because their outcomes do not give significant amount of data and because statistically homogeneous data could not be obtained.

When the contents of the studies under the title of “others” are reviewed, it was seen that the scores of those studies constitute a heterogeneous group, that sometimes the tests and evaluations with the same name have different contents. Indeed, although these tests and evaluations generally intended different measurements, since they do not have a systematic structure, their qualitative and quantitative analyses could not be performed in the study. In all of the studies handled, it is reported that at least 1 child has achieved basic sound awareness thanks to the use of evaluation tools. Considering the variety in number of participants, basic sound awareness is observed in different numbers in all studies.

4.5. Academic Skills and Outcomes of Non-Audiometric Tests

In this section, the tests used in the studies which are not including the audiometric evaluations were used. QOL (Quality of Life) evaluation tools were not included in this section.

Until the study of Colleti et al. (2008) academic skills and non-audiological evaluations are seen not to be used separately. For the first time in the study of Colleti et al. (2008), non-audiological cognitive skills were presented together with audiological findings. In that study, LIPS-R test battery was used. This battery ensures non-verbal measurement of cognitive skills. In this study, it is emphasized that 9 children have shown statistically significant improvement. However, cognitive meaning of these scores have not been discussed. Following that study, Eisenberg et al. (2008) and Sennaroğlu et al. (2009) have used non-audiometric tests in their studies.

Eisenberg et al. (2008) have assessed behavioral and mental development of the child by KABC-II (Kaufman Assessment Battery for Children), Vineland-II (Vineland Adaptive Behavior Scales), BASC-II (Behavior Assessment System for Children) assessment tools. Accordingly, the child involved has completed 3 of 4 sub-tests in KABC-II assessment and got 93 points which is a moderate value. In Vineland-II assessment, the child got overall 74 8low score whereas in BASC-II no significant unconformity was found.

Sennaroğlu et al. (2009), have used Denver Developmental Screening Test II. Stanford-Binet Intelligence Scale assessment and tests in their studies. According to these assessments, intelligence test results of 5 children were Normal (90-109 IQ), of two children were Normal Dull (80-89 IQ) and of one child was Borderline (70 -79 IQ). Regarding the developmental conditions, 6 children were reported as showing normal development (other than language development), two children showing retarded development at various levels and one child showing advanced retarded development.

Couloigner et al. (2014) report that one child could not attend to school full time after ABI and one child had to start school one year later and has difficulties at the school.

Besides, Yucel et al. (2015) have reported that all children (n:36) in their study have attended individualized training program and 8 of them who are gravely hearing disabled attend school. Also, they emphasize that, 17 of them have education in mainstream schools with inclusive education.

5. Discussion

Before reviewing the outcomes of the studies, we should review the participant profiles. Thus, we can see from which profile the answers of research questions are shaped. Afterwards, the results of the study outcomes can be reviewed more clearly.

In the studies included in this systematic review (see Table 2), age range of the participating children (n: 328) is very wide. The children are from different categories from babyhood (9 months) to adolescence (17 years). In the studies (n:36), 0-17 year old children are assessed as pediatric participants without dividing them into different categories. It reveals the fact that, the test and assessment tools which do not require compulsory age interval are applied to quite varied age intervals.

Although some tests (such as ITMAIS/MAIS) have different versions for different age intervals, the ones such as CAP, MUSS and Ling 6 Test, which are used frequently in studies do not have different applications/versions for age groups. In the studies involving participants from a wide age interval, the researchers tend to apply different test batteries to different age groups. For this reason, multiple assessment and test tools have been used in some of the studies (n: 26) included in the systematic review. Also, no tendency was determined towards using more assessment tools in the studies with more participants. As an example, in the study conducted by Colletti et al. (2013) involving 21 participants, only CAP assessment tool has been used whereas in the single case study of Eisenberg et al. (2008), 3 different assessment tools (ITMAIS, Ling 6 test and ESP) were used.

Besides the wider age interval, the gender distribution of 106 female (32%) and 93 male (29%) among the participants, shows that there isn't a significant gender difference within the participant group. However, the ratio of participants whose gender is not determined shouldn't be ignored. Genders of 129 (39%) of the participants of all studies are not indicated. In the light of these information, it can be said that the researchers have not establish research designs by which they can assess "gender" differences. Moreover, in the literature, there aren't other studies asserting that gender differences affect the outcomes of ABI users (adults and children).

The indications of the participating children that have caused ABI implementation are basically divided into two groups (NF2 and non-tumor). However, non-tumor group is divided into multiple groups within itself. According to the findings of included studies, in 26 (8%) of the children, MF' origin indications are observed while the reason of 302 (92%) children was non-tumor. Non-tumor group constitutes majority of the participants taking place in the reviewed studies.

It is supposed that this situation has started with the first non-tumor surgery in 2001 and accelerated with the approval of FDA the ABI implantation for non-tumor patients (Colletti et al, 2001; Goffi-Gomez et al, 2012). Moreover, the fact that anomalies and disorders in the non-tumor group are within a wide spectrum have extended the area of application of implants that bring hearing capability such as ABI and CI.

In the studies included in this systematic review, 258 children with 15 different non-tumor disorder anomalies were applied ABI and 8 children's (although being in the non-tumor group) anomalies or disorders have not been indicated. However, because indications and their level of affecting outcomes are not the subject of this study, indication-outcome relationship has not been investigated which causes a restriction to fort his study.

Surgery approaches are among the information which are overemphasized in the studies. The surgical method was indication in all studies (n: 31, 86%) except 5 (14%). Among then, the mostly used methods are retrosigmoid, translabyrinthine, retrolabyrinthine, suboccipital in order. The reason why surgery approaches take place, 86% (n:31) of studies as an important information is the fact that the overwhelming majority of studies are medically focused and most of the studies have been conducted by medical area researchers. Among the ones that take place within this systematic review, there isn't any study which establishes a direct relationship between the surgery approach and outcomes. It is thought that the surgery approaches and their possible affects on the outcomes are better to be examine on the basis of medical area.

All of the participating children suffer hearing loss due to various indications (ABI is not applied to individuals without hearing loss). According to the information obtained from studies, 64 children have one or more (other than hearing impairment) additional disabilities. In 16 studies, it is indicated that the children do not have

additional disability and in 11 studies, no information is given about disability. In 9 studies, the children were reported to have additional disabilities. Generally, in 25 studies (69%) additional disability is given as an information.

The basic reason for giving such information is supposed as possible causal relationship between the existence of disabilities and outcomes of assessments. The most significant support to this assumption comes from the studies indicating that the participants do not have additional disabilities. For example, among the exclusion criteria of Puram et al. (2016) the following statement takes place: “Lack of potential for spoken language development. This will be considered the case when evidence of the following exists: Severe psychomotor retardation, autism, cerebral palsy, or developmental delays beyond speech that would preclude usage of the device and oral educational development”

Accordingly, additional disabilities are assumed to potentially prevent language and speech development/training. As another example, the children in the study of Wilkinson et al. (2017) have been reported as not exhibiting retarded development cognitively or developmentally. In that study, the reason for excluding children with additional disabilities was stated as the fact that additional disabilities impair the collaboration between researchers and the child (Wilkinson et al, 2017).

As it can be understood from these examples, the concern related to the claim that additional disabilities might developmentally affect other areas outweighs. In a sense, not including the children without additional disabilities in the studies causes the research outcomes to address only a limited area. Not including the additional disabilities in the studies assuming that they will affect the assessment outcomes or research process might cause issues such as selection bias dominate the studies. In this study, not any broad review have been made about how additional disability affects audiological outcomes which constitutes a restriction for this systematic review. On the other hand, researching how multiple disabilities affect outcomes looks like a h-good opportunity for future studies.

When we reviewed the audiological and non-audiological outcomes, the mostly preferred ones among the systematically organized assessment tools were determined as CAP, ITMAIS/MAIS, MUSS and Ling 6 test in order This outcome is

similar to the outcomes of previous studies (Noij et al, 2015). Non-systematic assessment tools were combined under “other” category.

If these assessment tools were existed as a single tool, they would be the mostly preferred tool instead of CAP. Among these studies, the studies such as open and closed set recognition, identification, detection/recognition/discrimination/identification of instrumental sounds, fundamental phonetic sound detection, speech detection/perception,/discrimination, FC SS (suprasegmental four-choice test), FC (four-choice spondee test), Vowel (vowel/consonant/vowel presentation), open-set sound identification/recognition of bisyllable words/sentence recognition, and speech tracking, environmental sounds and onomatopoeic sounds, Open-set discrimination of polysyllabic number and similar take place.

There isn't enough information in all studies about the contents of these assessment tools which leads to the fact that some tests are applied to children without standardization.

None of the assessment tools used in studies have been specifically designed for ABI users. Some of the tests are the assessment tools which were previously used on CI users (Martines, Martines, Ballacchino & Salvago, 2013; Liu et al, 2014; Beadle et al, 2005). Looking at this perspective, there aren't custom assessment tools to be used for the assessment of the audiological outcomes of children with ABI. However, in certain patient groups, ABI implantation was demonstrated to lead better outcomes than CI implantation or other interventions (Peyre et al, 2013; Colletti et al, 2004c; Ramsden, Khwaja, Green, O'driscoll, & Mawman, 2005, Vesseur et al, 2018). However, there aren't advanced tools for ABI users to understand that if this difference is caused by the properties of devices or interventions.

When we look at CAP scores as the mostly used assessment, it is seen that the children can distinguish speech sounds. Because, 4 points basically shows that the participant can distinguish the speech sounds. The points below 4 indicates that the children are distributed between the levels of children's not having awareness of environmental sounds and distinguishing environmental sounds.

Here, the point that should be overemphasized is that the average CAP scores were obtained from studies which have outcomes clear enough to be analyzed. Also, there are studies which do not present individual score of the participants but presents

children's scores generally. For example, in the study of Yucel et al. (2015), it is reported that majority of the participants have got 5 or above points and the points are distributed between 1 and 7 but individual points are not given. As it can be in the results section, the CAP scores that can be analyzed distribute between 1,3 and 4,4. This distribution shows that the participants' average is at least at the level of awareness of environmental sounds. Based on the analyzable CAP scores, it can be said that the users are at the level of awareness in the average.

ITMAIS/MAIS scores must be assessed differently. ITMAIS/MAIS basically is a 40 questions survey completed by the hearing implant user or his/her parents. Due to its design, ITMAIS deals with the existence and levels of observable behaviors. When we look at from this perspective, ITMAIS/MAIS scores involve subjective values.

Moreover, it is a clearer situation for MUSS. When we compare the ITMAIS/MAIS and MUSS applications on children with cochlear implant, there are questions especially regarding feasibility and efficiency of MUSS (Allum, Greisiger, Straubhaar & Carpenter, 2000, Bosco et al, 2005; Pinto, Lacerda, & Porto, 2008).

For this reason, based on the systematic review, it does not seem possible to say that any test alone can be enough to assess the audiological and language development of children with ABI.

Education and Beyond

In the 36 articles reviewed, there isn't any study which only assesses educational and other developmental areas except the audiological outcomes. This situation clearly demonstrates that audiological assessment of children with ABI takes precedence over assessments in other developmental areas. Besides, there are deep quantitative and qualitative differences between the studies in the medical area and those in other developmental areas. Among the studies reviewed, only 5 studies have included non-audiological assessments and only two of them have made situation assessment directly for academic skills (Couloigner et al, 2014; Yucel et al, 2015). In the studies of Eisenberg et al. (2008) and Senaroğlu et al. (2009), there are assistive methods used to support explanations about cognitive capabilities, audiological capabilities and developments of the participating children. It is more

correct to say that this situation is caused by medical perspective. In a sense, we can say that the reason of this perspective is the concerns that ABI is influenced by cognitive factors. That is why the researchers have tried to prove that the outcomes obtained are independent from cognitive factors. Lastly, when we look at the study of Colleti et al. (2008), the researchers finally state that ensuring audiological rehabilitation affects the development of cognitive skills and thus highlighted the importance of the development of audiological skills.

As we can see up to this point, it is clear that the academic skills and the situation in other developmental areas are overlooked in the studies, which reveals a research gap needed to be filled. The educators and families strives for the education of their children every day. The experts in the medical are overlook the process, the education, for which children between 3 weeks and 144 months they follow up (see Table 2) spend most of their time and they focus on a limited area. On the other hand, there are resources stating that multidisciplinary development of implanted children shall be better for their development (De Raeve, 2010).

As a result, based on this systematic review we can say that the studies about children with ABI focus on a limited area and that, assessment and follow up efforts in other areas are not satisfactory.

6. Conclusion

Initially, the basic purpose of this systematic review was to find out which assessment tools are used to assess children with ABI and to determine their gains and then, to find out at what level their skills in academic and other cognitive areas were investigated. However, in the course of review studies, it was seen that majority of the studies in the literature were made considering mostly audiological outcomes. There was a big difference between the number of studies which assess educational and other areas and those which only present audiological gains. From the positive side, educators have a wide research opportunity whereas on the other hand, they have passed over the researches for children with ABI.

When we look at the outcomes of systematic review, a multidisciplinary point of review is definitely necessary to fill this gap. Because, in order to consider ABI implantation as successful, clearly multiple parameters should be evaluated. At this point, the educators must take more responsibility and collect and handle more data about the education of ABI implanted children. In this way, sound discussions can be made about whether or not children need ABI implantations and whether or not verbal-auditory skills are the only alternative for communication. If the number alternatives can be increased in the special education world, it can be ensured that both parents and the disabled individuals live without relying on a single model. In the scene obtained by this systematic review, it can be seen that the hearing loss caused by certain disorders and anomalies are perceived (in the medical area) as deficiencies which should certainly be recovered. Furthermore, the motivational factors leading to a risky surgery at the brainstem of a 9 months infant (Colletti et al, 2014) should be deeply examined. This study cannot present a perspective in this regard.

On the other hand, the studies included in the systematic review reveal that majority of the participants exhibit progress at basic audiological levels (awareness of environmental sounds, facing towards sound etc.). None of the studies in the systematic review advices not applying ABI or implies any negative information about its efficiency. Indeed, rather than demonstrating that ABIs are miraculous devices,

this situation points out another limitation of this study. Unfortunately, the studies which were not published or which could not find the opportunity to be published could not be included in this systematic review. When we look from this perspective, maybe those studies which have not been published because including negative results could not be included. In order to eliminate this concern, more studies must be conducted in educational and other developmental areas and ABI outcomes must be assessed from different perspectives.

It was observed that, the studies included in this systematic review have not followed a specific system in the audiological assessment of children but frequently used some assessment tools. However, because the existing data is heterogeneous, no deduction could be made about the efficiency of assessment methods utilized in the ABI studies. It is clear that, systematic assessment tools whose validity and consistency have been proved on ABI implanted children are needed for healthier assessments. When we look at the outcomes of studies, the variety of assessment batteries may lead to negative rather than positive results. The assessment tools which can address ABI implanted children from a broader perspective may prevent potential manipulations.

Another conclusion of this systematic review is that, educators do almost no study in this area. Children with ABI seem to be completely left for medical area. The challenges experienced by educators and the obstacles met by children or the parents' opinion are not given place in the literature. Therefore, based on this systematic review it is not possible to reach any conclusion about whether or not ABI implanted children have taken inclusive education, whether or not their audiological skills affect their academic skills or whether or not special education methods have been developed. However, we can say that there are good research opportunities in this field.

Consequently, studies on ABI implanted individuals should be conducted multidisciplinary and the educator must focus in this field. Also, clearly more systematic and specific assessment tools are needed in this field. The question of whether or not ABIs are miraculous devices may only be answered after eliminating these deficiencies.

7. References

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8. Appendices*

*This section has been taken from researcher's previous dissertation draft.

8.1. Brief History of ABIs

On 24 May 1979, Dr. William House and his team successfully implanted the first Auditory Brainstem Implant (ABI) at the House Ear Institute (House & Hitselberger, 2001). A few years before the successful implantation of the first ABI, the first successful cochlear implant operation had been performed. This was a revolutionary step in the restoration of hearing loss. However, this revolutionary success achieved with cochlear implants was met with a lackluster reaction from certain academic circles with very high expectations because of certain technical difficulties. The first cochlear implant models had a single channel. This meant that it had deficiencies in transmitting sounds from the 8th cochlear to the auditory nerve. Moreover, recipients of a cochlear implant had to be completely free of inner ear anomalies or any problems in the area where the electric signals would be transmitted from the electrodes to the auditory nerve. Therefore, for the implant to work, the "auditory nerve" had to be healthy enough to transmit the signals created by the implant to the brain. Otherwise, a single-channel implant would not work.

The relationship between the human body and electrical stimuli had been known since the trailblazing studies of Luigi Galvani (1773-1831) (Whittaker, 1960). However, the idea advanced by Djourno and Eyries in 1957 that hearing impairments may be corrected by electrical stimulation to the inner ear would only find practical application in the 1960 when progress in technology allowed the practical implementation of the theoretical idea (Møller 2006). A few years after the publication of Djourno and Eyries' study, the first cochlear implant was successfully used (Møller 2006). The next step was the discovery that similar methods could be successfully used to correct hearing loss where cochlear implants failed. William House and his team realized that the fact that the auditory nerve could be electrically stimulated could also be applied to similar structures (House & Hitselberger, 2001). The most suitable test group for their hypothesis was sufferers of Neurofibromatosis type 2 (NF2), as, as it will be mentioned later on, a tumor or several tumors situated in NF2 patients can permanently damage the auditory nerves. Another factor was the fact that the surgical procedures used to remove tumors from NF2 patients could largely and permanently damage the auditory structure of a patient. House and Hitselberger theorized that as Neurofibromatosis Type 2 suffered permanent hearing loss after the removal of their vestibular schwannomas by the trans-labyrinthine craniotomy, also because of the nature of the disease itself, cochlear implants developed before ABI would be insufficient in correcting the hearing loss of such patients (Cervera-Paz & Manrique, 2007). House and Hitselberger's approach to ABI was more focused the inadequacy of the working principle of cochlear implants on the anatomical and physiological components of NF2 patients and people with similar conditions. Shortly, NF2 patients either lack the anatomical structure needed to carry electrical signals up through the auditory nerve to other auditory areas of the brain or these areas are damaged beyond repair and are unable to transmit incoming electrical signals. Therefore, a more suitable auditory

structure was found for NF2 patients that would allow them to transmit the signals from the implant: cochlear nucleus.

The patient in the first case of House and Hitselberger (2001) had had a tumor removed from his left ear in 1964 by the trans-labyrinthine craniotomy method. Another tumor was found in the right ear, which heard normally, of the same patient 12 years later. After the necessary steps were taken, an ABI implant was successfully put in place to correct his hearing loss for the first time in history. Around the same period, cochlear implants (CI) were also yielding successful results. However, both first CIs and the newly designed ABIs were single channel implants. As will be further discussed later, this created problems, especially in the transmission of the sound of speaking. The first multi- channel ABI was developed in the early 1990s. 21- channel ABI implants made by the Nucleus Company were used successfully for 10 patients between 1992 and 1995 (Laszig, Marangos, Sollmann, Ramsden, Fraysse, Lenarz, Rask-Andersen, Bredberg, Sterkers, Manrique, and Nevison, 1997). According to Coletti et al. (2005) state that the 21- channel Nucleus-22 model was used until 1999, when the same company released Nucleus 24.

After the suitable area for the implant was determined, the implants, today named ABI, were used for a long time on NF2 patients. Although released around the same time, ABIs did not produce the same audiological outcomes as cochlear implants (Lim, Lenarz, Joseph, Battmer, Patrick & Lenarz, 2008; Moore & Shannon, 2009). This suggests that there are differences between these implants and that these differences influence their effects on the anatomical auditory areas stimulated by the implants (Fayad, Otto, Shannon & Brackmann, 2008).

Before we move on to the differences between cochlear implants and ABIs, we shall take a quick look how are they working and which pieces have they:

8.2. Auditory Brainstem Implants

ABIs are composed of two main parts:

1. The external part
2. The internal part

The external part includes a microphone that captures the incoming sounds, a sound processor to process the captured sound and a magnetic transmitter that keeps the implant stuck to the patient's head and transmits the incoming sounds to the internal part of the implant. The internal part, on the other hand, is made up of a section where the incoming sounds are transferred to the electrode and the electrode that is place to the brain stem. The essential difference between ABI implants and cochlear implants is the electrodes. In recent years, penetrating electrodes were developed to make the

most of the tonotopic structure of the brain stem area where ABI implants are positioned. Unlike electrodes that touch the surface of the brain stem, the ABI electrodes are placed inside the brain stem. Therefore, they look more like nails than other, surface electrodes. While this technology, comparatively more recent than surface electrodes, have not exceeded predictions of studies, they had more or less the same audiological outcomes as surface electrodes (Otto, Shannon, Wilkinson, Hitselberger, McCreery, Moore & Brackmann, 2008).

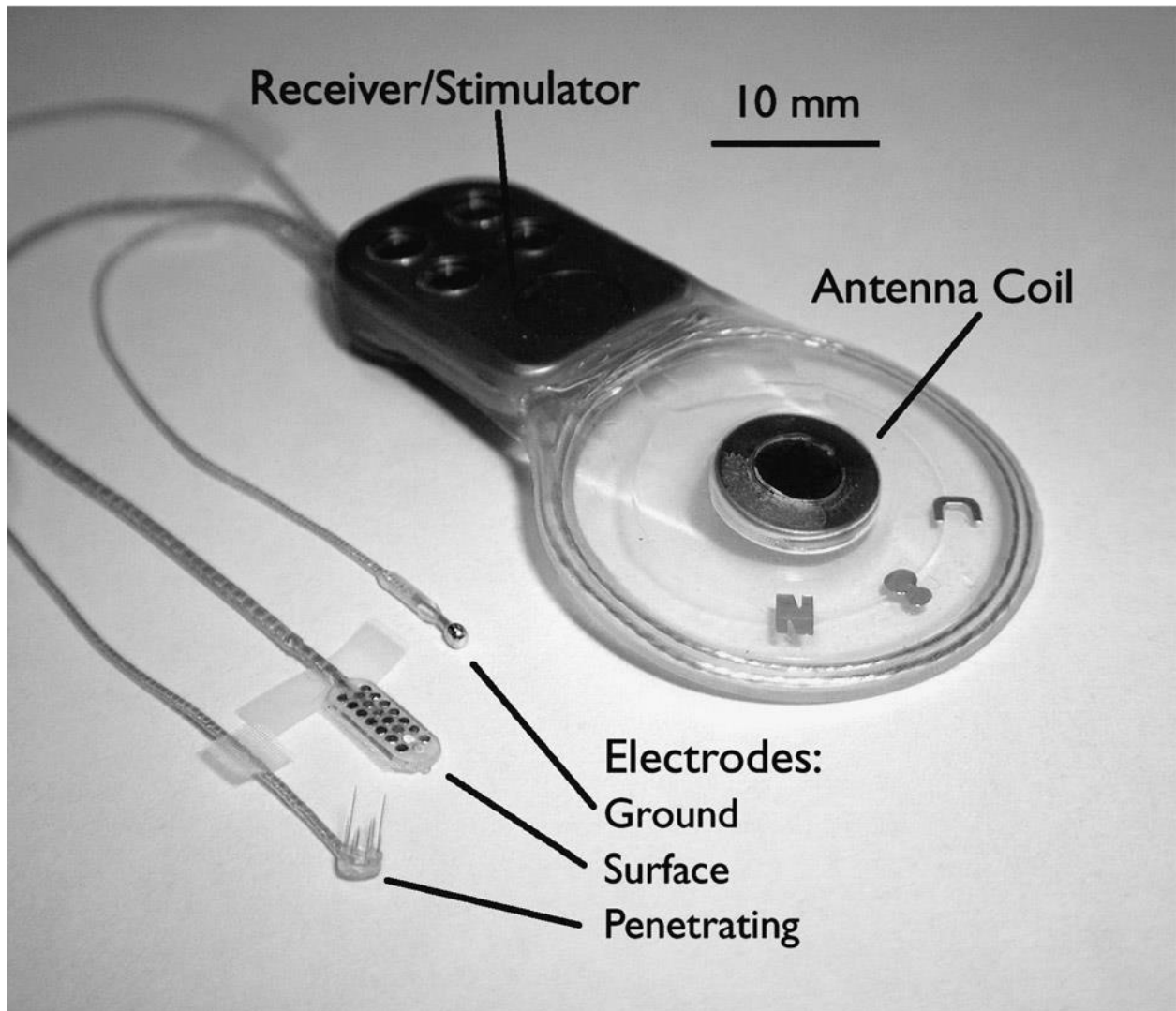


Fig. 1: ABI internal parts and different types of electrodes (Otto et al., 2008) (Ground electrodes are used to reduce electromagnetic parasites and are not transmitters)

Today, ABI manufacturers invariably use similar external and internal parts. This generally means that implant manufacturers make similar devices but have differences in more specific terms such as sound processing algorithms, processors, voice detection strategies and number of electrodes. For example, while Cochlear, the first company to develop the multi- channel implant for ABIs, currently has 21 electrodes in their modern ABIs, Medel ABI implants have 12 electrodes. (Fig. 2) Another ABI manufacturer, Neurelec, puts 15 electrodes on their implants. (Fig. 2)

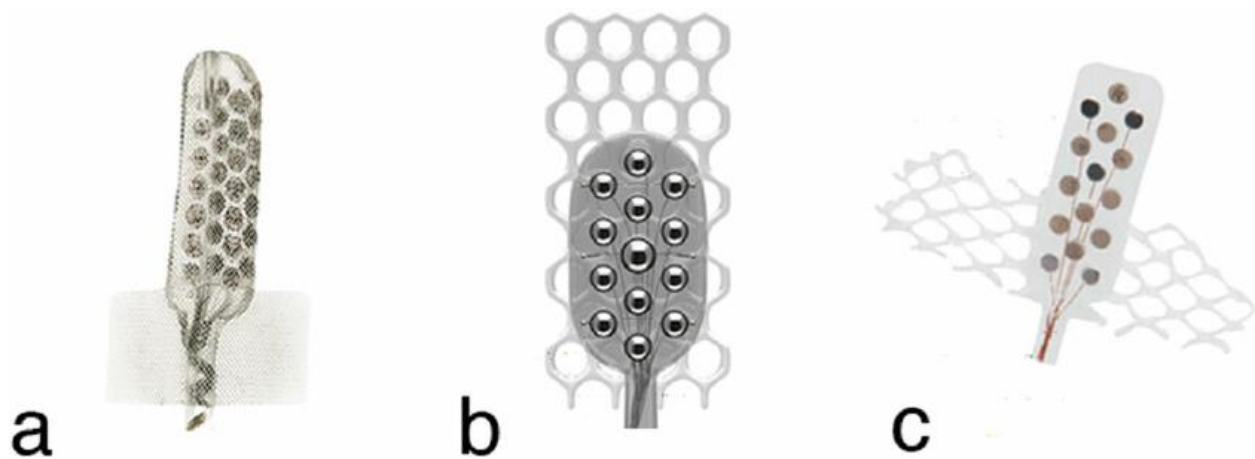


Fig. 2: Electrodes by manufacturer: a) Cochlear b) Medel c) Neurelec (Vincent, 2012)

Processors and sound processing algorithms used by 3 main ABI manufacturers in their implants are completely different from one another. Interestingly, these manufacturers do not develop specialized processors or sound processing algorithms for ABIs, instead opting to use processors and sound processing algorithms they use for their cochlear implants. This shows that essentially the manufacturers do not aim to increase output by following tailored strategies for ABIs, and choose to try and replicate the systems and strategies that made the cochlear implant strategy such a big success. While there has not been any research conducted on how this affects performance in ABI users, Schwartz and Wilkinson (2016) point to financial factors to explain why manufacturers do not develop specific systems and sound processing algorithms for ABIs. Schwartz and Wilkinson (2016) state that there are around 2000 ABI users in the world and that ABI devices are being manufactured as a matter of prestige and PR strategy. Therefore, it can be said that despite considerable advances in cochlear implant technology, manufacturers have not had any considerable developments when it comes to ABI technology.

8.2.1 How do ABI implants work?

In order to understand the working principle of ABIs, the working mechanism of their forerunners, the cochlear implants, need to be explained. This is because, as previously mentioned, ABI implants are transformed versions of cochlear implants in terms of their internal and external parts as well as their sound processing algorithms. As can be inferred from their name, cochlear implants work by an electrode lodged in the inner ear (cochlear) transmitting the electrical signals of the implant to the auditory nerve. As cochlear implants work directly through the inner ear, they bypass the outer ear and the tympanum and directly stimulate inner and outer hair cells that transform mechanical sound into electrical signals, therefore forming a kind of electrical sound simulation. It can be said that the main reason cochlear implants have very successful output is that they make use of the auditory imprints of the inner ear. This is why the electrodes of cochlear implants, as opposed to ABIs, have been designed to take the shape of the inner ear. Therefore, the electrodes can separately and locally stimulate hair cells responsible for transmitting tonotopically different frequencies of sound. The signal sent by the cochlear implant by stimulating inner hair cells reaches the auditory

cortex through the following path: The auditory nerve, the cochlear nucleus, the thalamus and the auditory cortex (Fig. 3) (Rauschecker, & Shannon, 2002).

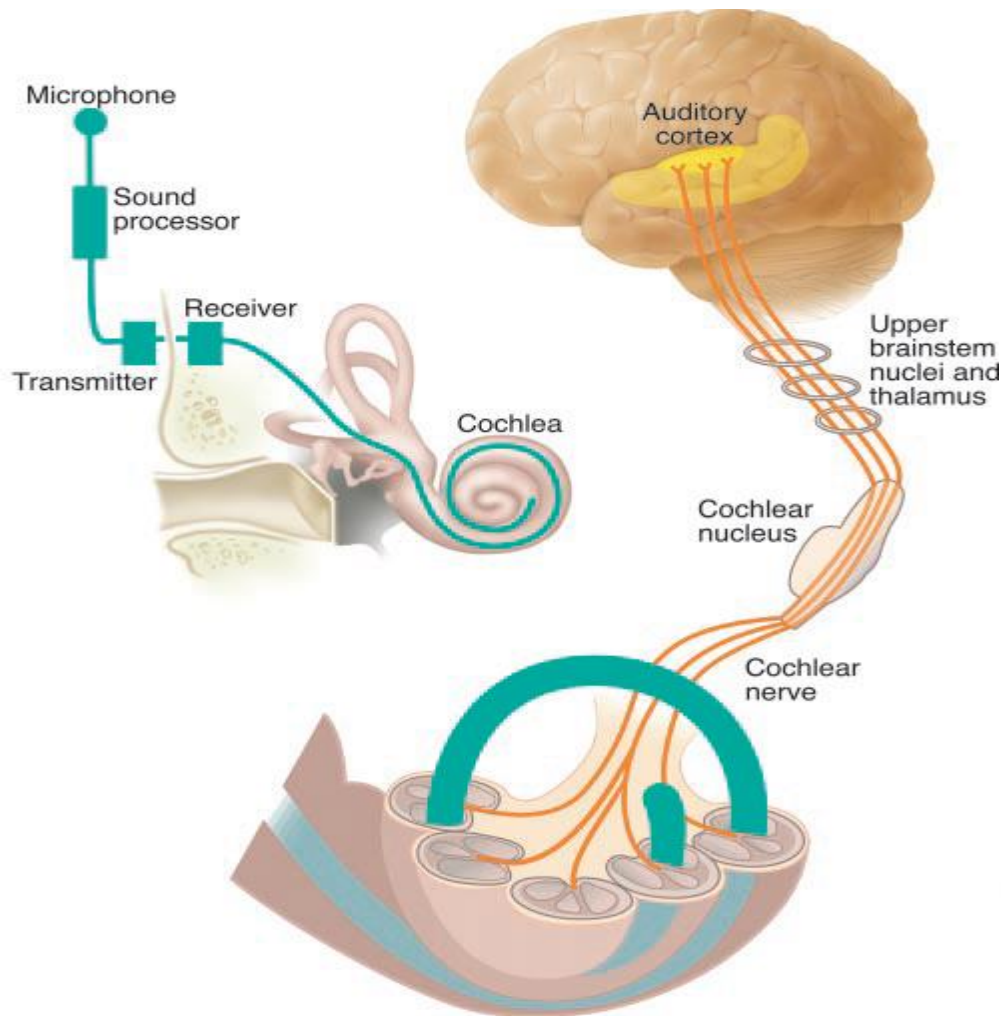


Fig. 3: Cochlear implants and their anatomical working principles (Rauschecker, & Shannon, 2002).

As it can be seen, cochlear implants make use of the auditory structure of the inner ear (the hair cells), therefore ensuring the transmission of the electrical signal to the brain in a tonotopically organized fashion (Guiraud, Besle, Arnold, Boyle, Giard, Bertrand, ... & Collet, 2007). The structures used by the cochlear implants are natural structures already used in individuals of sound hearing in everyday life. As an example, while inner hair cells are responsible for converting mechanical energy into electrical energy and transmitting it to the auditory nerve, the cochlear implant uses electrical energy to stimulate hair cells. Therefore, what cochlear implants essentially do is to simulate the working principle of a healthy ear. Especially children in their critical language acquisition period therefore show high levels of acclimatization to their cochlear implants following a neuroplasty (Moore, & Shannon, 2009). The difference between the working principle of the ABI implant and the cochlear implant, on the other hand, is where the electrode is placed and the electrical signal is transmitted. ABI implants bypass the entire structure of the ear and are connected directly to the brain stem area called the cochlear nucleus where the auditory nerve meets the nerve system.

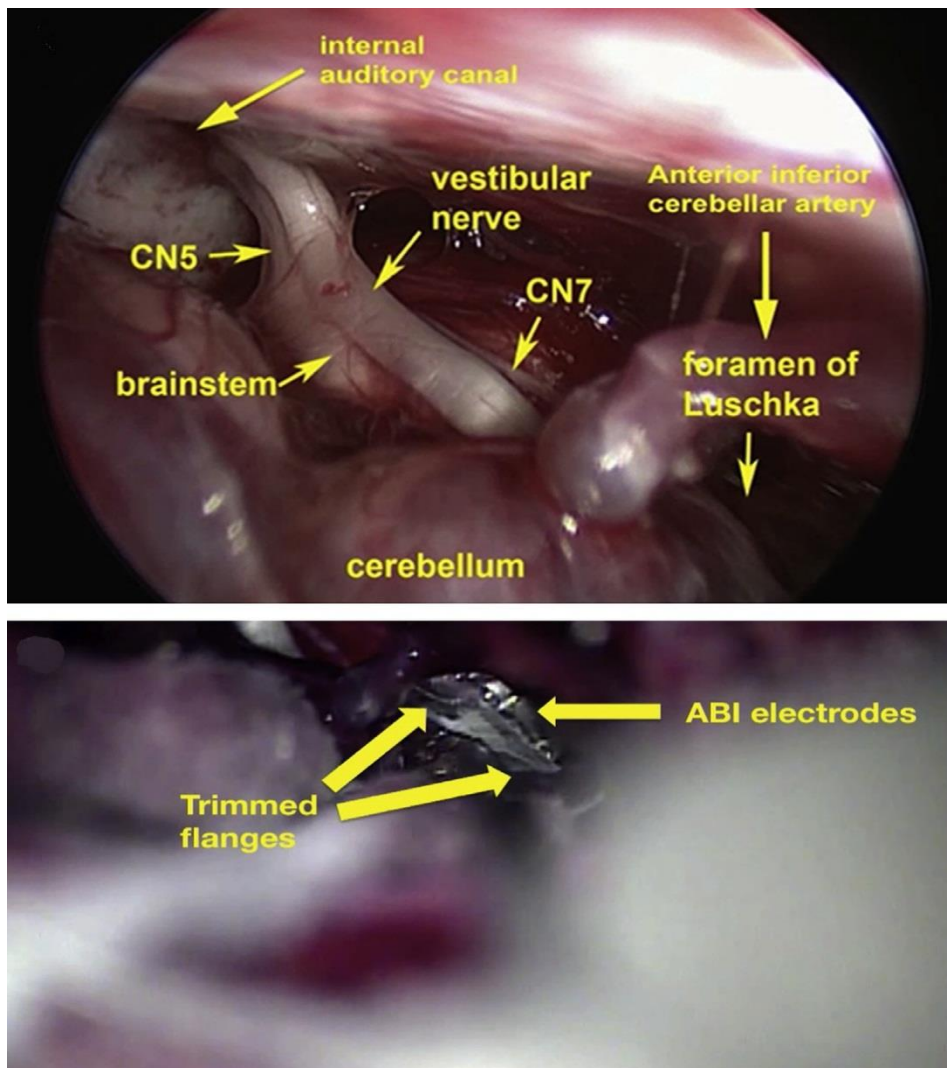


Fig. 4: The locations of the brain stem and the nearby anatomical areas where ABIs are positioned (Puram, & Lee, 2015).

Apart from where the electrodes are placed, the working principles of the two implants are very similar. Sound data gathered by the external part of the system is transmitted from the internal part to the external part, while the signal converter in the inner part forms electrical signals and transmits them to the electrode. As previously mentioned, the sound processing technology used by ABI is the same as the one used by cochlear implants. However, this technical similarity is not reflected in ABI outcomes and the conversion of surface electrodes into penetrating ones did not produce any significant improvements (Otto et al., 2008). This creates the impression that the difference in the zone stimulated by the electrode also affects the outcome. As Vincent (2012) has claimed, although ABIs work, we cannot say with any certainty how. One of the main reasons for this is the fact that we do not have a lot of information on how tonotopical organization works in human beings after the inner ear (as tonotopical research is usually conducted on primates and other mammals, most research about human beings is based on assumptions). (Humphries, Liebenthal, & Binder, 2010). In order to understand how ABI implants work and why they perform less well when compared to

cochlear implants, we need to better understand the anatomy of the area where ABI electrodes are placed.

8.3. Anatomical Approach to ABIs

ABI electrodes are placed in an area that is known as the cochlea nuclei or the cochlea nucleus. This area is also where the vestibulocochlea nerve, the auditory and balance, 8th cranial nerve, enters the brain stem (Komune, Yagmurlu, Matsuo, Miki, Abe, & Rhoton Jr, (2015). All acoustic data received from the ear is transmitted to this area and later transferred to the auditory cortex. The cochlea nucleus has a crucial position along with the structures that are around it. VII. Cranial nerve is very close to essential nervous system structures such as the pons and the medulla (Fig. 5) (Puram & Lee, 2015; Fayad et al., 2008)

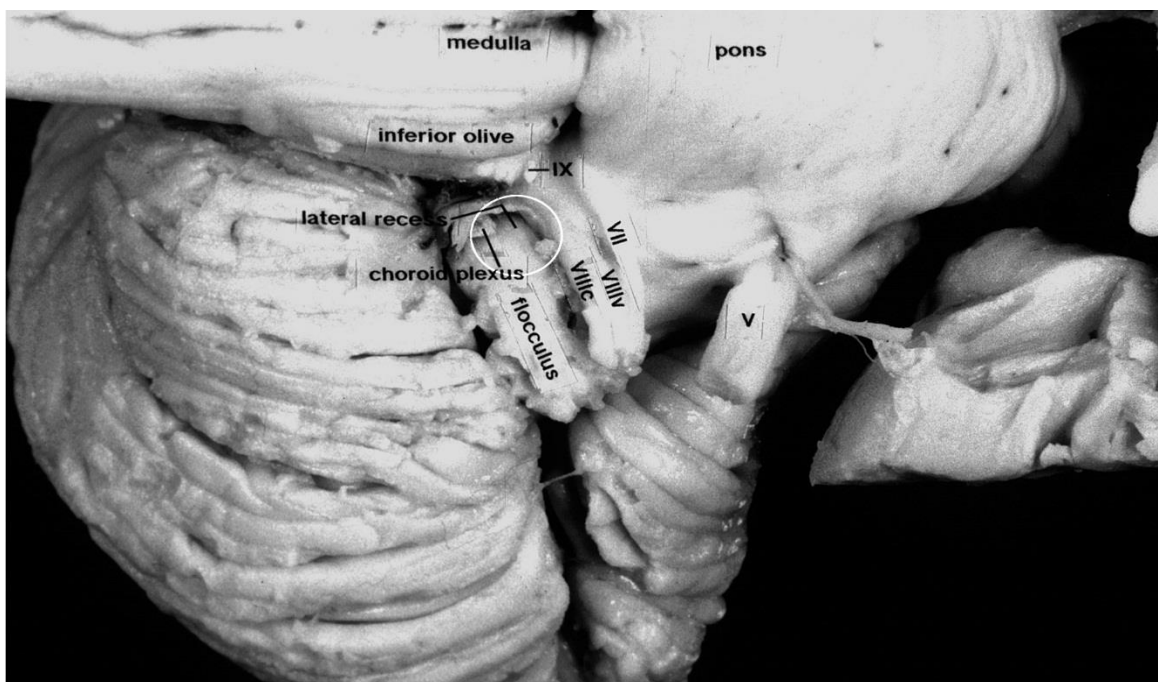


Fig. 5: The anatomical position of the Cochlea Nucleus (within the white circle) and the surrounding anatomical areas.

Another reason why this area is important is that it is prone to permanent damage when the auditory tumors of NF2 patients are removed through different surgical methods, therefore significantly affecting ABI performance (Colletti et al, 2000; Puram & Lee, 2015). In other words, the surgical method used on NF2 patients affect the outcome of ABIs. Moreover, as the cochlea nucleus includes several types of cells with the particular structure, certain difficulties are experiences when processing the signals sent by ABI (Vincent, 2012). The Cochlea Nucleus is made up of 4 main parts (Fig. 6b) (Terr, 1985):

- Dorsal Cochlea Nucleus (DCN)
- Ventral Cochlea Nucleus (VCN)
- Anteroventral cochlear nucleus (AVCN)

-Posteroventral Cochlear Nucleus (PVCN)

These parts also include at most 4 different types of cells (Fig. 6a) (Kiang, 1965):

-Fusiform

-Octopus

-Stellate

-Bushy

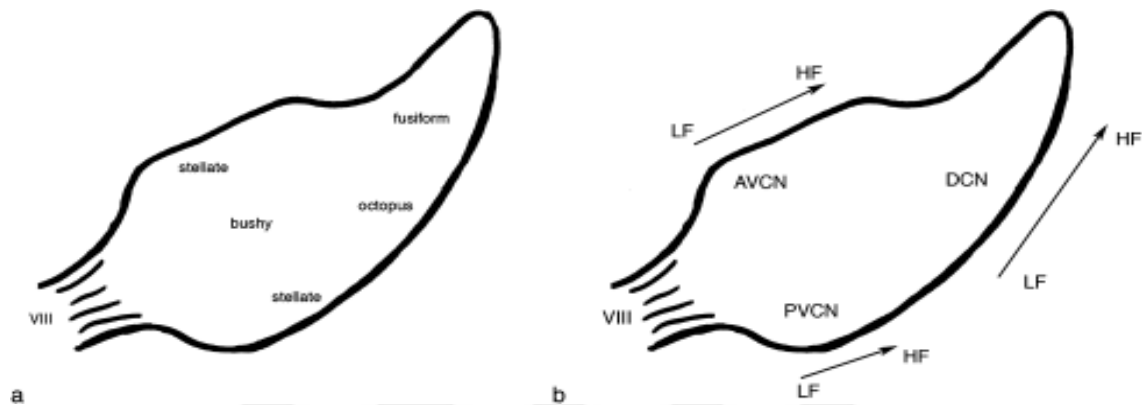


Fig. 6: Parts of the Cochlea nucleus and its cell types (Vincent, 2012)

Each type of cell in the cochlea nucleus fulfills different functions. For example, while the fusiform cells serve to pause the signal coming from the cochlea nucleus, the octopus cells allow the signal to pass. However, ABIs with surface electrodes are placed on a PVCN-DCN axis on the cochlea nucleus and are thus unable to stimulate electrically all the areas of the cochlea nucleus. Newly developed penetrating electrodes are, on the other hand, only placed along the PVCN-AVCN axis and thus cannot stimulate all the areas of the Cochlea Nucleus, just like the surface electrodes (Fig. 7). Although Rauschecker and Shannon (2002) claimed that it was possible for penetrating ABIs to provide better tonotopical stimulation, Otto et al. (2008) found that this did not meet expectations in terms of output.

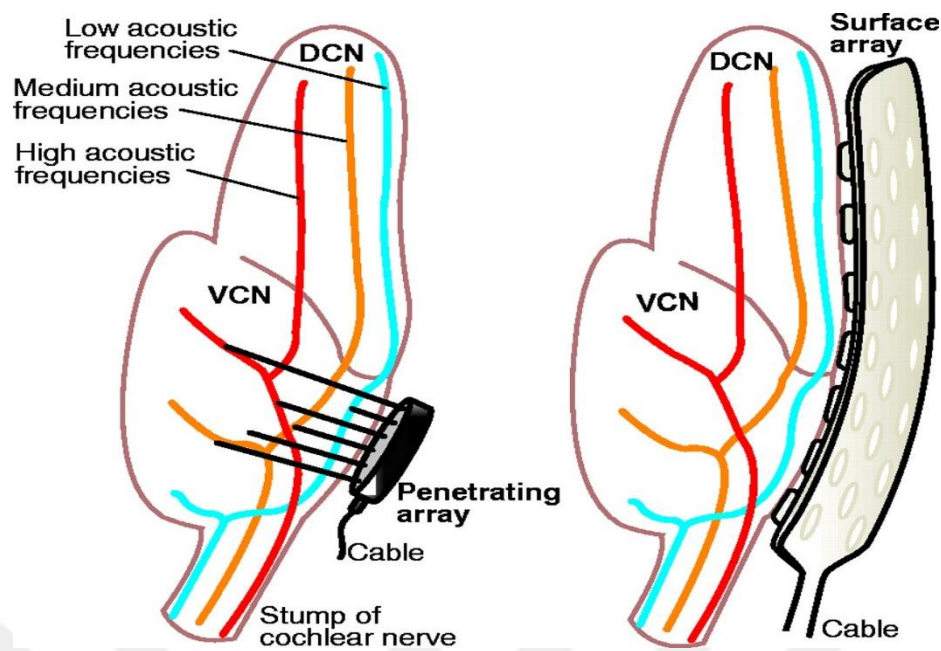


Fig. 7: Areas of the Cochlea Nucleus that can be stimulated by surface and penetrating electrode. (Rauschecker & Shannon, 2002)

As a result, although ABIs are similar to cochlear implants in terms of technical features and work in a similar fashion, they stimulate different anatomical areas. As previously explained, cochlear implants ensure better transmission of sound to the auditory cortex as they provide better tonotopical stimulation of the zone they stimulate through electrical signals. However, we do not have a clear knowledge of the level of stimulation ABIs provide to the cochlea nucleus area in tonotopical terms. Moreover, because of the particular structure of the cochlea nucleus, the outcome compared to cochlear implants remains weak despite the use of different electrodes. This means that different strategies are needed for patients requiring ABI implants. Additionally, the working principle of ABIs and the zones they stimulate through electrical signals affect not only the outcome but also results such as speech recognition. Colletti and Shannon (2005) state that the reason why individuals with ABIs underperform is the pathways used by ABIs in the transmission of signals. A similar argument may be advanced for audiological outcomes. A study conducted on the reactions of ABI users to human voices in their superior temporal sulcus (STS) and temporal voice areas (TVA) where human voices are processed showed that the activation in the TVA areas is superior to the activation of ABI users (Coez et al., 2009). The same study showed that the reaction of ABI users to auditory stimulants is stronger to the reactions of individuals with normal hearing abilities (Coez et al., 2009). This shows us that although ABI users process auditory stimulants, their visual zones are more active than their auditory zones thanks to cross-modal reorganization. In short, although signals reach the auditory areas in ABI users, they cannot be adequately processed.

Additionally, while ABIs are not suitable for every hearing-impaired individual, it is the only alternative in case of some diseases and anomalies to rehabilitate hearing. These diseases and anomalies may be grouped as follows:

8.4. ABI Treatable Indications

ABI implantations may be used for two main groups:

- 1-Individuals with postlingual hearing loss
- 2-Individuals with prelingual hearing loss

1-Individuals with postlingual hearing loss

Individuals within this group are further separated into two categories: 1.1. Patients with tumors caused by Neurofibromatosis type 2 and 1.2. nontumor patients (Sennaroglu&Ziyal, 2011).

1.1 Neurofibromatosis type 2 Patients

Neurofibromatosis type 2 (NF2) is an autosomal dominant genetic disease caused by mutations to the tumor-suppressing 22nd chromosome, therefore causing the body to present multiple neoplasias (Asthagiri, Parry, Butman, Kim, Tsilou, Zhuang, and Lonser, 2009). Although the frequency of its appearance is different all around the world (for example, 1 in each 33-40.000 births in the UK, 1 in 87.000 in Finland (Evans, Huson, Donnai, Neary, Blair, Newton, & Harris, 1992; Antinhenio, Sankila, Carpen, Pukkala, Sainio and Jääskeläinen, 2000), the world average is 1 in 60.000 (Evans, 2009). One of the most common symptoms of NF2, seen in 90-95% of all patients, is bilateral vestibular schwannoma. (Evans et al., 1992; Mautner et, Lindenau, Baser, Hazim, Tatagiba, Haase, Samii, Wais, and Pulst, 1996). Vestibular schwannomas are homogenic tumors on the VIII. cranial nerve that expand toward the brain stem. These tumors announce their presence by such symptoms as hearing loss and tinnitus, seen in 60% of all adult patients and 30% of all child sufferers (Maccollin and Mautner, 1998; Evans, Baser, O'reilly, Rowe, Gleeson, Saeed, King, Huson, Kerr, Thomas and Irving, 2005; Evans, Birch, and Ramsden 1999; Nunes and Maccollin, 2003). As NF2 patients suffer from hearing loss or damage to their auditory structure, especially on the VIII. cranial nerve during the surgical removal of vestibular schwannomas, cochlear implants do not produce positive results for NF2 patients (Colletti, Carner, Miorelli, Guida, Colletti and Fiorino, 2004a). Especially the trans-labyrinthine craniotomy method to remove vestibular schwannomas, while effective in preventing the formation of new tumors, NF2 patients cannot usually regain their hearing abilities as the surgical method causes permanent damage to the auditory areas (Colletti, Fiorino, Carner, Giabini, Sacchetto and Cumer, 2000; Wilkinson, Schwartz, Otto, and Brackmann, 2014; Ramsden Khwaja, Green, O'Driscoll, and Mawman 2005).

1.2 Non-tumor patients

Hearing loss problems of patients in this group are diverse as they are caused by different conditions of the middle and inner ear areas. Sennaroglu and Ziyal (2011) categorizes patients in this group as follows:

1.2.1 Cochlear Ossification

Some bacterial meningitis patients are left unsuitable for cochlear implants because of the effects of meningitis on their cochleas. As cochlear implants are placed in the scala tympani canal in the inner ear, permanent deformations in the area such as cochlear ossification make the placement of cochlear implants difficult. Moreover, it has been reported that cochlear implants to patients with bilateral loss do not provide the desired audiological outcomes (Sanna, Khrais, Guida & Falcioni, 2006). While it has been theorized that in cases of cochlear ossification, ABI implants would produce better and more satisfactory results (Bozorg Grayeli, Kalamarides, Bouccara, Ben Gamra, Ambert-Dahan, & Sterkers, 2007), a recent study on 9 patients with cochlear ossification implanted with ABI devices show that the outcome does not reach the expected level (Bayazit, Kosaner, Celenk, Somdas, Yilmaz, Altin, Cevizci, Yavuz & Ozluoglu, 2015).

1.2.2 Advanced Otosclerosis

Advanced otosclerosis is a term first used by House (1961, cited in Calmels, Viana, Wanna, Marx, James, Deguine, & Fraysse, 2007) to describe patients with an airway auditory sensitivity of 85 dB or more that suffer from long-term chronic otosclerosis. Hearing loss caused by advanced otosclerosis can be largely corrected by traditional hearing devices, surgical interventions to the stapes bone and also recently with cochlear implementation (Calmels et al., 2007). However, Sennaroglu and Ziyal (2011) state that the loss of bone minerals caused by advanced otosclerosis mean that the cochlear implant strongly stimulate the facial nerve and thus create problems in the placement of the cochlear implant electrode. This creates difficulties in the programming of the cochlear implant. It is thought that ABI implants are an alternative for patients who cannot use cochlear implants (Sennaroglu & Ziyal, 2011).

1.2.3 Other anomalies and diseases

Any loss of the transmission pathways between the cochlea and the cochlear nucleus cause hearing loss that cannot be corrected by cochlear implants. Especially in non-tumor cases with auditory nerve damage, ABI cochlear implants can produce better results (Colletti, Carner, Miorelli, Colletti, Guida, & Fiorino, (2004b). Moreover, it has been claimed that ABI implants may be an alternative for various cases where cochlear implantation do not produce the desired results (Sennaroglu & Ziyal, 2011). In 2014, A comparative study conducted on child patients suffering from hearing loss caused by

inadequacies in the auditory nerve showed that patients with ABI implants (n = 94) have a significant advantage over patients with cochlear implants (n = 443) in certain areas (open-set speech perception, acquiring verbal language competence etc.) (Colletti, L., Colletti, G., Mandalà, & Colletti, V., 2014).

2-Individuals with prelingual hearing loss

This group includes individuals that started suffering from hearing loss before the age of 2 as a result of birth anomalies of the inner ear. Sennaroglu and Ziyal (2011) determined the anomalies of such patients as follows: Complete labyrinthine aplasia, cochlear aplasia, cochlear aperture aplasia, IAC aplasia, and aplastic cochlear. Complete labyrinthine aplasia refers to the absence of inner ear structures (such as the membranous and osseous labyrinths) as a birth defect (Ozgen, Oguz, Atas, & Sennaroglu, 2009). For Jackler, Luxfor, and House (1987), cochlear aplasia refers to the complete absence of the cochlear. IAC aplasia, on the other hand, refers to the absence of the internal auditory canal where the auditory nerve travels along the temporal bone or an underdeveloped auditory nerve that cannot fulfill its function (Baik, Yu, Kim, K. S., & Kim, G. H. 2008). Cochlea aperture aplasia, on the other hand, refers to the lack of the point where the auditory nerve enters the cochlea or the cases where the aperture is completely covered by cochlea tissue (Glastonbury, Davidson, Harnsberger, Butler, Kertesz, & Shelton, 2002). Hearing loss caused by all these anomalies and malformations cannot be corrected by cochlear implantation as they constitute an obstacle to the proper functioning of the cochlear. More importantly, according to initial research, ABI implantations on individuals with inner ear malformations do not produce positive audiological outcomes (Sennaroglu et al., 2009).

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