

SAUDI SOCIETY OF SPEECH- LANGUAGE PATHOLOGY AND AUDIOLOGY NEWSLETTER (SSSPAN)



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Family-Centered Practice

The professions of Audiology and Speech-Language Pathology have advanced from Client-Centered to Family-Centered Practice. A Family may include parents, guardians, siblings, spouses and caregivers (ASHA). Family-centered practices include offering more active roles for families in the planning, implementing, Interpreting and decision making in service delivery (ASHA, 2008).

Audiologists and Speech-Language Pathologists (SLPs), in their daily practice, deal with different varieties of clients of a wide age range (from infancy to elderly). In family-centered practice, families play an essential role in all aspects of the service (ASHA). Thus the family is an integral part in prevention, assessment and therapy of almost all hearing, balance, communication and swallowing disorders. According to a national survey, 80% of SLPs indicated they gave parents activities to complete at home (Pappas et. Al., 2008).

Pediatric language disorders, for example, demonstrate the importance of family engagement in prevention, assessment and therapy. The language disorder can be prevented or at least reduced in severity if the family visited an SLP at an early age and applied his/her instructions to help stimulate the child's language. When a language disorder is suspected, the main source for information about the child's general and language development is the family. Language assessment procedures that rely on reports from parents such as Receptive-Expressive Emergent Language Test Third Edition (REEL-3) (Bzoch, League & Brown, 2003) and The JISH Arabic Communicative Developmental Inventory (Dashash & Alsafi, 2014) are evident examples of family involvement in the process of assessment.



SSSPA VISION

To be the representative of Hearing, Balance, Speech-language and Swallowing Professionals nationally and internationally.

SSSPA MISSION

SSSPA's mission is to support scientific and professional interests, and provide efficient services for professionals in the science and treatment of communication, swallowing, hearing and vestibular disorders both inside and outside the Kingdom. It also encourages their cooperation to enhance the capabilities of Saudi individuals with hearing, balance, communication or swallowing disorders. In addition to influencing health and education policies to support the profession and the provision of specialized services

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Language therapy is useless without family involvement. The typical language therapy session is based on specific aims that are targeted during the session and carried over by the family at home. Similarly, this is applicable to other hearing and communication disorders.

It is essential for a clinician to be skillful in dealing with the family to achieve a more effective intervention. This can be demonstrated by different examples discussed in the next paragraphs. When obtaining case history information it is important to ask about family factors that might have an affect on the process of assessment and intervention such as the structure of the family, the person who takes care most of the child, other family members present at home (i.e. grandparents, aunts...). The clinician needs to figure out the extent to which a parent is willing to participate in the intervention process. This is specifically true for some parents who think that intervention process is clinician-centered and their role as a family is very minimal. When communicating with family members, the language used should be simple and clear. Education of parent and their culture should be considered when exchanging information with them. The session usually terminates with tasks that the family is requested to perform. These requests need to be realistic and consider the time and skills of family member (s). In short, the family-centered practice is essential and more effective to the clinician, the client and his/her family.

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Low Expectation is as Bad as Unrealistically Higher Expectation in Cochlear Implant Practice



Alfarghal Mohamad, MD, AuD

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I have been extensively working with cochlear implant candidates, recipients and their families for more than 8 years. As any clinician in the field, we have been taught that unrealistically higher expectations is one of the worst scenarios, and sometimes considered a relative contraindication for cochlear implantation, yet no body warned us about the drawbacks of very low expectations. In my experience, I have seen about 10 cases, where very low expectations have negatively affected the outcome of cochlear implantation.

(We are very happy and satisfied that our son/daughter can hear well, and talks well or achieved great progress in language development), a very relaxing message, which has a narcotic effect on the rehabilitative team. Parents and families of profoundly deaf children sometimes have this serious low expectation, problem, they give very positive and satisfactory feedback for any little progress, they keep comparing their child performance before and after the implant, rather than comparing it to the average hearing age peers or to the normal hearing peers.

Clinicians could be deceived with pleasant feedback and became reluctant to continue improvements in the recipient's map, this could waste very precious time during the critical period of speech and language acquisition. Validated outcome measures such as validated questionnaires, age appropriate normative data charts, distance specific Ling Six sounds testing with normal voice, cochlear implant aided audiometric thresholds should be the routine monitoring tools rather than subjective parental feedback .

Families should be counseled not only about unrealistic expectations but also about the drawback of very low expectation. Reasonably high expectation would better motivate the parents and keep them highly engaged in the cochlear implant rehabilitation process.

“ The sky is our limit ”



Nora Fahad Al-Sudairi, MSc

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THE SSSPA SCIENTIFIC WORKSHOPS AND CLUB MEETINGS

The Saudi Society of Speech Language Pathology and Audiology (SSSPA) has established the “**SSSPA Scientific Club Meetings and Workshops**” to promote interest in advancement of evidence based clinical practice, to enhance knowledge and understanding in the field in Saudi Arabia, to become one of the primary venues for audiologists and communication/swallowing clinicians to present their new work to their colleagues and to promote learning about new research, tools and assessment/therapy techniques that might be relevant to their clinical practice. Its main goal is to bring the audiology and communication/swallowing clinicians' community together and to provide opportunities for them to interact. In case you missed the workshops or the meetings, below is a summary.

The 1st Advanced Audiology Cochlear Implant Course (AACI)

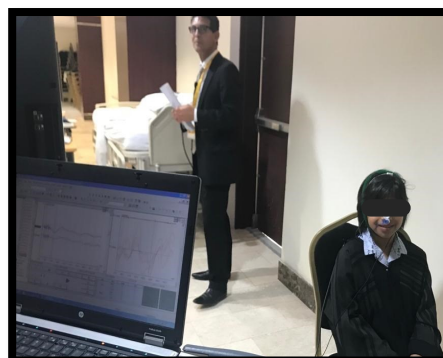
Was held on 13th-15th of March 2018. The course was organized by King Abdullah Ear Specialist Center at King Abdulaziz University Hospital with collaboration with SSSPA. Attendance was limited to 20 Audiologists only due to the hands-on nature of the course and to ensure positive interactions. The course ran from 8:00 a.m. to 5:00 p.m. and was accredited 30 CME credit hours by the Saudi Commission for Health Specialties.

The workshop provided Audiologist with highly specialized and advanced information in addition to hands on practical training with Cochlear Implants. It was designed to provide in-depth knowledge and practical training on neurophysiological testing with Cochlear implants. As well as present Audiologists

with the most recent research and technical information to enhance evidence-based practice when dealing with complex cochlear implant cases. The course was presented by: Prof. Andy J. Beynon, Dr. Barry Nevison, Dr. Shaza Saleh, Mr. Mohammed Shadid and Eng. Hassan Yalcouy.

It is worth mentioning that this course, is the first advanced course of its kind to be held in Saudi Arabia that specifically targets senior and experienced Audiologists who work with Cochlear implants.

Dr. Hattan AL-Khayari concluded the meeting by expressing his appreciation to all speakers and volunteers and to Cochlear company for sponsoring the course.



The Best Practices in Aphasia Assessment & Intervention: A Review Workshop:

Was accredited 6 CME/PD credit hours by the Saudi Commission for Health Specialties (SCFHS) (No. 18000046588).

Dr. Areej AlAsseri and Dr. Yaser AlSa'bi concluded the workshop by expressing their appreciation to the speakers and to the volunteers from Dar Al-Hekma University, who helped organizing the meeting and handed them certificates.



Dr Areej Asseri

Best Practices in Aphasia Assessment & Intervention: A Review Workshop

On Saturday, the 5th of September 2018, the “**Best Practices in Aphasia Assessment & Intervention: A Review Workshop**” took place at Dar Al-Hekma University, Jeddah. It was attended by 21 SLPs, interns and students. The workshop was from 9:00 a.m. to 4:00 p.m.

Opening speech was presented by Dr. Sameera Duhaithem, Dean of school of Health, behavioral sciences and education. Followed by the first lecture titled “**Aphasia Classification**” was presented by both Dr. Areej AlAsseri, PhD, CCC- SLP, Assistant professor, Provost, and Dr. Yaser AlSa'bi, PhD, CCC- SLP, Assistant Professor, Director of Master of Speech-Language Pathology Program.

The second lecture titled “**Best Practices in Aphasia Assessment**” which consisted of two parts, the first part was presented by Dr. Yaser AlSa'bi, PhD, CCC- SLP, Assistant Professor, Director of Master of Speech-Language Pathology Program.

Which then was followed by the second part which was presented by Dr. Areej AlAsseri, PhD, CCC- SLP, Assistant professor, Provost.

Then was followed by “**Best Practices in Aphasia Intervention**” which contained two parts, the first part was presented by Dr. Areej AlAsseri, PhD, CCC- SLP, Assistant professor, Provost and, the second part was presented by Dr. Yaser AlSa'bi, PhD, CCC - SLP, Assistant Professor, Director of Master of Speech-Language Pathology Program.



Dr Yaser AlSabi



Pre-test

Auditory Verbal Therapy Course

The “**Auditory Verbal Therapy Course**” took place at King Abdullah Ear Specialist center, Riyadh from Tuesday to Thursday 25th- 27th of September 2018. It was attended by total of 60 communication/ swallowing clinicians-SLPs, Audiologists, Interns and students. The course was from 8:30 a.m. to 5:00 p.m. and was accredited 30 CME/PD credit hours by the Saudi Commission for Health Specialties (No. 18000046820).

The course highlighted the principles of **Auditory Verbal Therapy, Cochlear Rehabilitation Materials, Children with Hearing Loss and Literacy and, Auditory Verbal Therapy for Children in School**. Which were presented by Mr. Warren Estabrooks, M.ED., Dip.Ed. Deaf, LSLs Cert. AVT., Dr. Nadia Abdulhaq, PhD, AuD. And Miss. Beauty Hariz.

Prof. Abdulrahman Hager concluded the meeting by expressing his appreciation to the speakers and to the volunteers from the King Abdullah Ear Specialist center and Cochlear company who helped organizing the meeting and handed them certificates.

Since SSSPA is going green, certificates of attendance were sent to them via e-mails.

Tracheostomy and Speaking Valve – Lecture



The ***“Tracheostomy and Speaking Valve – Lecture”*** took place at Prince Ahmed Bin Abdulaziz Auditorium – Security Forces Hospital – Riyadh on Saturday 29th of September 2018. It was attended by 80 communication/ swallowing junior clinicians- SLPs, interns and students. The course was from 10 a.m. to 12:00 p.m. and was accredited 3 CME/PD credit hours by the Saudi Commission for Health Specialties (No.18000046804).

The lecture titled “Tracheostomy and Speaking Valve” was presented by Mrs. Maha Al-Delaigan, Msc, CCC – SLP at King Faisal Specialist Hospital and Research Center.

Mr. Talal AlZurgi (SSSPA President) concluded the meeting by expressing his appreciation to the speakers and to the volunteers from the Speech and Hearing Student Club- (King Saud University) who helped organizing the meeting and handed them certificates.

The 1st Cochlear Implant Fitting Fundamentals Course

Since SSSPA is going green, certificates of attendance were sent to them via e-mails.

Was held on 28th - 30th of October 2018. The course was organized by King Abdullah Ear Specialist Center at King Abdulaziz University Hospital with collaboration with SSSPA. Attendance was limited to 35 Audiologists and speech language professionals interested in Cochlear Implants because of the hands-on nature of the course. The course ran from 8:00 a.m. to 5:00 p.m. and was accredited 30 CME credit hours by the Saudi Commission for Health Specialties.

The workshop provided Audiologist/SLPs with fundamentals of Cochlear Implant programming and follow-up, speech processing strategies, candidacy issues, objective measures and troubleshooting in addition to hands on practical training with Cochlear Implants. The course was presented by: Dr. Florian Feichtner, Dr. Shaza Saleh, Dr. Medhat Yousef and Mr. Juergen Danzer in addition to an interactive troubleshooting session presented by two of Medel’s specialist team.

Dr Hattan AL-Khayari concluded the meeting by expressing his appreciation to all speakers and volunteers and to Medel company for sponsoring the course.



The SSSPA Fifth Scientific Club Meeting

Dr. Abdulrahman AlAkeel (SSSPA Vice President) concluded the meeting by expressing his appreciation to the speakers, JISH for hosting the meeting and to the student volunteers from Dar AlHikma University- Speech and Hearing program who helped organizing the meeting. Dr. Abdulrahman also emphasized on the fact that SSSPA is for all the members and should be participating more in all the activities.



The **"SSSPA fifth scientific club meeting"** took place at the Jeddah Institute for Speech and Hearing JISH, Jeddah on Thursday, 8th of November 2018. Dr. Nahla Dashash, the JISH Clinical Director and SSSPA board member opened the meeting by welcoming the guest and highlighting the role of SSSPA for SLP's and Audiologist. The meeting was attended by 28 SLPs, Audiologists and students. The meeting was from 4:00 to 7:00 p.m. and was accredited 3 CME/PD credit hours by the Saudi Commission for Health Specialties (No. 18000047781).

The first lecture titled **"Application of Lee-Silverman Voice Treatment Program on Saudi Lady with Parkinson's Disease: Case Study"**, which was followed by the discussion of the

paper **"Review of the Effect of LSVT on improving vocal quality for patients with Parkinson's disease and related neurological disorders."** Both of which were presented by Ms. Hala Abuliya, MSc-CFY-Speech -Language Pathologist Jeddah Institute for Speech and Hearing JISH.

The second lecture titled **"Single sided deafness and unilateral hearing loss, an update"** which was followed by the discussion of the paper **"Single sided deafness and unilateral hearing loss"**. Both of which were presented by Dr. Nisreen Naita, CCCA- AAAP, Consultant Audiologist, American Board of Audiology, King Faisal Specialist Hospital, Jeddah, Saudi Arabia.



SSSPA's First Debate Competition

Mr. Saleh AlSuwailim concluded the workshop by expressing his appreciation to the speaker and to the volunteers from Princess Nourah University and King Saud University, who helped organizing the debate competition and handed them certificates.

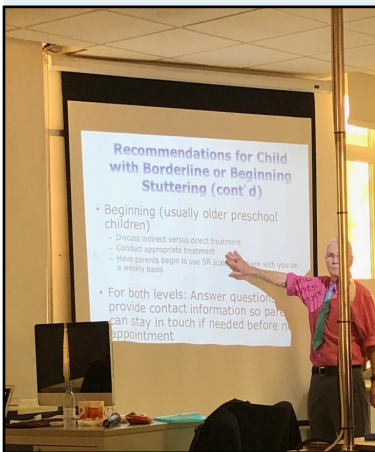
On Saturday, the 3rd of November 2018, the **"SSSPA's First Debate Competition"** took place at King Abdulaziz University Hospital, Riyadh. It was attended by 23 SLPs and Audiologist, junior clinicians, interns and students. The workshop was from 1:00 p.m. to 4:00 p.m. and was accredited 3 CME/PD credit hours by the Saudi Commission for Health Specialties No. (18000048375).

Opening speech and lecture titled **"Introduction to Debate"** was presented by Monera AlAbdullatif, SLP intern. Then the first topic of the debate competition was presented **"Standardized language assessment (such as PLS) can be applicable with children with autism"**. Followed by the second topic of the debate competition was presented **"PECS is useful for verbal output in children with autism"** and were judged by Mr. Saleh AlSuwailim (SLP), Miss Ala Elrashid Hussien Osman (Psychologist), Mr. Ahmed Gomaa (Special Education), Miss. Elham AlQathmi (SLP) and Miss Ghada AlSanae (Mother).

Stuttering Workshop, Jeddah

Stuttering Workshops:

Since SSSPA is going green, certificates of attendance were sent to them via e-mails.



Stuttering Workshop, Riyadh

On Saturday and Sunday, 1st and 2nd of December 2018, the “**Stuttering: An Integrated Approach to Its Assessment and Treatment**” took place at Security Forces Hospital, Riyadh. It was presented by Dr. Barry Guitar and Mr. Goncalo Leal. It was attended by 55 SLPs, interns and students. The workshop was a two-day workshop. The first day was from 7:30 am to 5:00 pm and the second day was from 8:00 a.m. to 5:00 p.m. and was accredited 16 CME/PD credit hours by the Saudi Commission for Health Specialties (No. 18000048614).

Opening and closing speech was presented by Dr. Abdulrahman Ibrahim Al-Akeel, Ph.D.-SLP, Vice-President of the SSSPA.

Mona Jawad AlAmeer, Audiologist.

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King Fahad Specialist Hospital- Dammam.

January, 2018.

Etiology of Single- Sided- Deafness Among Patients at King Fahad Specialist Hospital-Dammam

Introduction:

Single- sided deafness (SSD) is defined as severe to profound Sensorineural hearing loss in one ear and with normal or near to normal hearing in the other ear (Kimura et al., 2015) & (Christopher et al., 2014). Scientists consider SSD as the severe degree of unilateral hearing loss, and can cause reduced school performance among children and affect the social or emotional activity between adults (Usami et al., 2017).

Prevalence and incidence of SSD until now is unknown, however, the prevalence has been estimated at 3 to 6% of the population (Ross et al., 2010).

One of the major problems that the SSD patient faces is the head shadow affect, where sound coming from the side of the

deprived ear will drop in head shadow and will be detected by the better ear (Cire et al., 2012). Studies suggested that up to 10-16 dB HL attenuation of the signal occurs due to head shadow affect (Vermeire et al., 2009). Low frequencies contain long wavelength and bend easily around the head to reach the opposite ear (good ear). While high frequency (HF) sound, which has short wavelength, is more likely to be attenuated and therefore, cannot reach opposite ear. In addition some consonant sounds are located within HF; therefore, SSD patients reported difficulty in speech discrimination and understanding (Cire et al., 2012).

Objective:

The aim of this study is to report the main etiology of SSD among KFSH-D patients.

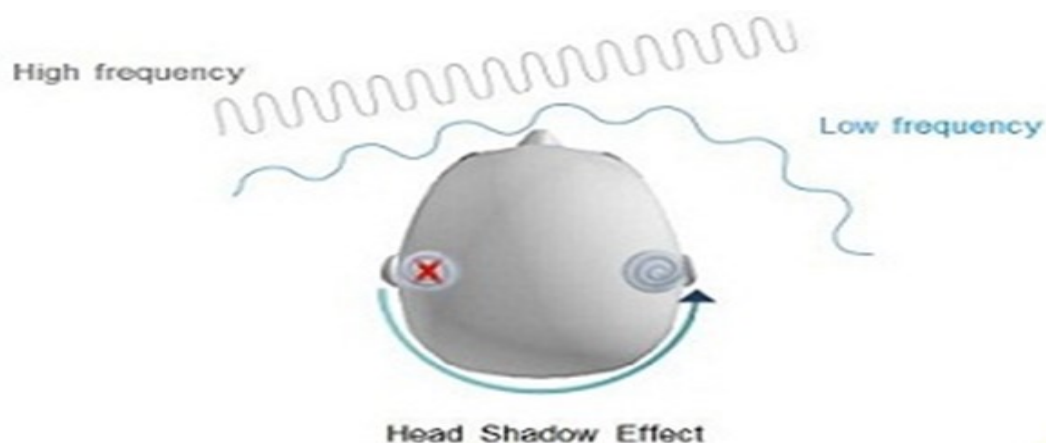


Figure (1)

Spatial hearing or localization which is the ability to identify the sound coming from "which side". The consequences of SSD includes the loss of that ability. Caused by the inability to detect interaural time difference and interaural level difference. For the binaural hearing person, sounds will be localized within 360 degrees on every side of the head (Cire et al., 2012). Human ears could differentiate within millisecond between signal stimuli when coming from one side or the other, this mechanism (ITD) occurs at low frequencies (LF) <1500 Hz (Cire, 2012) & (Blauert, 1997). On the other hand, ILD occurs at HF above 1500 Hz, which

means that the sound is louder at nearest ear than the other (Blauert, 1997).

SSD occurs due to various causes, including acoustic neuroma or vestibular schwannoma, head trauma, cochlear nerve deficiency, sudden sensorineural hearing loss (idiopathic), congenital SSD, Mumps virus and Meniere's disease (Usami et al., 2017) & (Cire et al., 2012). Through advanced technology implant, patients with SSD may improve a higher quality of life (Usami et al., 2017).

Intervention depending on the etiology of SSD will be considered as a first step toward appropriate treatment (Usami et al., 2017). For instance, when the cause sited within the inner ear (cochlea), a cochlear implant would provide positive outcome. However, if the etiology found within the auditory nerve or higher (central lesion), the cochlear implant will not provide positive outcomes (Usami et al., 2017).

Methods:

A retrospective study, data collected from medical files of patients who visited the ENT clinic for general examination and have Magnetic resonance imaging (MRI) or Computed tomography (CT) scans. All patients included were diagnosed after Audiological assessments at KFSH-D during the period of December, 2012 until December, 2017.

The total number of patients who were diagnosed with SSD was 33 and age ranged from 8 to 63 years.

Inclusion Criteria:

Pure Tone Audiometry (PTA) criteria: the non-functional ear hearing thresholds average for the frequencies 0.5, 1, 2 and 4

kHz should be ≥ 70 dB HL. Whereas the good ear (contralateral side) hearing thresholds average must be ≤ 30 dB HL. The interaural threshold difference between ears must be equal to or above 40 dB HL.

Only patients who had soft copy report audiogram saved through the GSI software were included.

Exclusion Criteria:

The patient who did not meet the criteria were excluded.

Result:

Etiology	Patients' Numbers	Percentage
Congenital SSD	8	24.2%
Vestibular Schwannoma	6	18%
Head Trauma	6	18%
Unknown Etiology	4	12%
Cholesteatoma	2	6%
Sudden SSD	2	6%
Acute Lymphocytic leukemia post chemo-therapy	1	3%
Perilymphatic fistula	1	3%
Labyrinth Ossification	1	3%
Jugular foramen tomure	1	3%
Cogan's syndrome	1	3%

Discussion:

In this study, the most common cause of SSD was congenital, 2 cases with Michel Aplasia (total absence of inner ear structure/no cochlear and vestibular organs) and were determined via CT scan. It is a rare inner ear malformation, that occurs in about 1% of cochlear bony abnormality (Yiin et al., 2011). Other 3 congenital cases had unremarkable CT or MRI whilst 2 cases had not done. Many papers reported cochlear nerve deficiency as the most common etiology of SSD in a Chinese Study 40% of SSD cases were attributed to CND (Usami et al., 2017). On the other hand, in our study SSD due to CND was not found, this could be due to the small number of SSD but could also be an indication that CND is not a common cause for SSD in Saudi Arabia.

Vestibular schwannoma was the second most common cause for SSD. This study showed that all Vestibular schwannoma cases were within the cerebellopontine angle lesion.

In this study, all SSD patients with Vestibular schwannoma had acquired progressive unilateral hearing loss (5 out of 6) except for one case who had sudden hearing loss which was pre-removal surgery.

Studies reported a positive correlation between the tumor size and the degree of hearing loss, as such large tumors would cause greater hearing loss but not always, however some small tumors might cause significant hearing loss (Pinna et al., 2012).

Head trauma which was associated with temporal bone fractures/ or labyrinthine concussion without fracture was also the second main causes of SSD. In our study out of the six head trauma cases, one had a transverse temporal fracture, two had no fracture, one had a longitudinal temporal fracture, one had Labyrinth Ossification and one case did not have a written CT report.

Transverse temporal fracture frequently caused SNHL according to several studies, while longitudinal temporal fracture caused conductive hearing loss (Schuknecht, 1956). However, longitudinal temporal fracture was also reported to cause SNHL (Maradi et al., 2017).

The complication of chronic otitis media with cholesteatoma is proven. The reason could be that the bacterial toxins leak from the round window to the inner ear which ends up with sensorineural hearing loss (Viswanatha et al., 2014). Two cases of SSD were associated with cholesteatoma. One case had SSD after petrosectomy for congenital Cholesteatoma and the 2nd case with acquired cholesteatoma.

Sudden single sided deafness was reported in 2 adult patients. One of the patients had SSSD post renal transplant, the other case acquired SSD after waking up with no other issues (both had normal radiological tests). No clear correlation between SSD and kidney transplant has been proven. Even though, one study suggested that high levels of tacrolimus (drug taken after organ transplant to reduce immune system) might be associated with sudden hearing loss. Two reported cases had sudden HL after kidney transplant which were associated with elevated levels of tacrolimus but the hearing loss was bilateral (Gulleroglu et al., 2013).

Post chemo-therapy for Acute lymphocytic leukemia was an unexpected etiology. In this study, an 8 year old patient noticed right ear hearing loss after 4 months post chemo-therapy. The patient also had history of head trauma since 6 years pre leukemia with no baseline hearing test but the CT scans showed normal temporal bone. It's hard to differentiate the cause of the SSD among these factors in this young patient.

On the subject of chemotherapy, studies proved that chemo-therapy with cisplatin can cause asymmetrical hearing loss instead of symmetrical HL. Cisplatin does

not always lead to symmetrical HL (Schmidt, 2008). Authors had some theories to explain why asymmetrical HL: first, because the blood flow is asymmetrical between ears; second, some studies suggest due to supracochlear issues and third theory refers to a cortical outcome. However, further research is necessary. (AAA, 2008).

A SSD has been reported in a case of Chronic Myeloid Leukemia (Hsu et al., 2004). Leukemia is considered as one of the etiologies of acute unilateral hearing loss (Harada et al., 2000).

Considering Hsu's study, the patient with SSD due to acute lymphocytic leukemia post chemo therapy, leukemia is the probable cause of SSD. However, additional investigation is needed to confirm the etiology and to exclude other etiologies. Moreover, otologist should keep in mind the possible association between sudden SSD and hematologic disorders such as leukemia so they would not be missed along with other different possibilities such as Vestibular schwannoma (Harada et al., 2000). Other SSD etiologies included perilymphatic fistula, followed by Cogan's syndrome (Autoimmune disorder), Jugular Foramen Schwannoma and finally, Labyrinth Ossification which appeared through CT, without history of infection or head trauma.

Regarding the intervention, Cross and Baha can be considered as alternative hearing solutions (Kimura, 2015). This should overcome the head shadow effect however would not restore binaural hearing and spatial listening. Some schools suggested that any kind of bone conduction aid is an optimal treatment option for all etiologies of SSD (Tahir, 2016) while others suggest cochlear implantation (e.g. Arndt et al., 2011).

Conclusion:

Single sided deafness is a serious medical problem which interferes with the patient's communication ability. Knowing the etiology of single sided deafness is the first step toward the provision of successful intervention. Moreover, it can draw attention regarding threatening medical issues which are associated with SSD.

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The Implication of Huntington Disease on Individuals from Speech, Language and Swallowing Perspectives in Saudi Arabia.

Abstract:

Huntington's disease (HD) is described as a genetic disorder caused by a mutation in the cytosine-adenine-guanine and has been described as a genetic disorder that affects speech, language and swallowing aspects. Depending on the level of the disease, individuals with HD may present difficulties in speech, language and swallowing. It is well documented that individuals with HD may display problems in speech, language and swallowing. Speech problems may include signs of hyperkinetic dysarthria due to non-controlled choric movements. Language problems may be demonstrated by the use of shorter and simpler statements,

and in some cases difficulties in tasks that require complex cognitive processing. Also, swallowing problems include dysphagia. Therefore, effective speech-language intervention is fundamental to improve the quality of life of individuals with HD and contribute to their communicative wellbeing. The purpose of this article is to present these difficulties and to shed light on some treatment strategies which may help overcoming these difficulties as well as to discuss studies related to HD in Saudi Arabia.

Keywords: speech, language, Huntington, swallowing, Saudi Arabia.

Introduction:

Huntington's chorea or San Vito's disease (Bhattacharyya, 2016), or Huntington's disease (HD) can be defined as a "common inherited neurodegenerative disorder which is characterized by cognitive, emotional deficits and uncontrolled excessive motor movements" (Chan, Walker, & Robertson, 2007). It is caused by mutation in the number of repetitions of the Cytosine-Adenine-Guanine (CAG) nucleotide sequence (Neylan, 2003). The average survival of the persons with HD varies between 15 and 20 years.

Huntington disease (HD) can be considered as "neurodegenerative disorder" which affects many aspects of life, such as motor and cognitive aspects (van Duijn, Kingma, & van der Mast, 2007). Huntington Disease (HD) is usually characterized by a distinguishing feature; chorea movements. The chorea movements occur due to loss of control on muscle movements in different areas, such as face, neck and oral cavity (Arango-Lasprilla, Iglesias-Dorado, & Lopera, 2003). It also may affect muscles involved in swallowing and communication.

Only few studies have reported the existence of HD among Arab families in Middle Eastern countries, including Saudi Arabia. This may be because many cases may have not been reported in Saudi Arabia. The prevalence of HD in Saudi Arabia is still vague; however, first-hand reports have claimed that the prevalence of HD in Middle Eastern Arabian countries varies between 3 to 4 cases per 100,000 (Scrimgeour, 2009).

Huntington Disease is a unique neurodegenerative disease which has three levels; in the first or initial level, an individual with HD presents with small changes in the coordination of involuntary movements; moreover, difficulty in solving problems (Meza-Escobar, Orozco, Takeuchi, Ariza, & Pachajoa, 2014), depressed mood and impulsivity behaviors may also be expected at this level (van, 2010).

In the second or intermediate level, an individual with HD presents with moderate choric movements, with variable speech and swallowing deficits. In addition, the cognitive aspect is gradually deteriorates with the appearance of disorientation and short-term memory loss (Ho & Hocaoglu, 2011).

In the third or late level, an individual with HD shows severe choric movements accompanied with muscular rigidity and swallowing problems. Weight loss, walking ability and talk ability are affected at this level, which may result in the individuals being dependent on others (Velasco-García, Cobeta, Martín, Alonso-Navarro, & Jiménez-Jiménez, 2011).

Impacts on Speech Perspective:

Since HD affects muscle movements and that since the speech mechanism involves the contribution of many muscles, individuals with HD usually present with speech difficulties, such as hyperkinetic dysarthria (Hamilton et al., 2012). In other words, HD affects respiration, phonation, articulation, resonance and prosody. This means the reduction of verbal communication occurring during the initial level of the disorder and may in some cases, lead to mutism at the third level (Kaploun, Saxman, Wasserman, & Marder, 2011).

Individuals with HD usually show greater effort during breathing (Hartelius, Carlstedt, Ytterberg, Lillvik, & Laakso, 2003). Moreover, breathing difficulties may appear unpredictably accompanied by sudden inspiration or forced expiration during connected speech. This may affect other aspects involved in the speech mechanism. Voice may also be affected in individuals with HD. Individuals with HD may showed perceived tense (Hamilton et al., 2012), harsh (Vogel, Shirbin, Churchyard, &

Stout, 2012), choppy or hoarse, vocalization with irregular tone fluctuations. These changes occur due to the involuntary contractions of the vocal folds. Phonation time reduction is also expected in HD cases due to insufficiency of airflow (Kaploun et al., 2011). Moreover, the resonance aspect can be affected, although this depends on the affected muscles; hypernasality due to defects in soft palate muscles is a clear example of resonance defects in HD cases.

Huntington disease (HD) also affects articulation; individuals with HD usually show distortion in sound production at vowel and consonant level (Rusz, Saft, Shlegel, Hoffman, & Skodda, 2014). Moreover, inappropriate pauses within words, sound prolongation and a slow speech are com

mon features in HD cases (Hartelius, Jonsson, Rickeberg, & Laakso, 2010).

Impacts on Language Perspective:

Huntington disease (HD) impacts can be extended to language perspective at comprehension and expression level.

At the *language comprehension level*, individuals with HD may present with difficulties in passive voice sentences, (Saldert, Fors, Ströberg, & Hartelius, 2010); moreover, lexical-semantic manipulation problems may appear in individuals with HD (Chenery, Copland & Murdoch, 2002). Individuals with HD may show difficulties in the executive functions of attention, short-term memory and inability in planning activities (Dayalu & Albin, 2015).

At the *expressive language level*, individuals with HD may present with reduction in sentence length; moreover, the sentence structure in individuals with HD tends to be simple with syntactic errors (Murray & Lenz, 2001). Grammatical errors such as the omission of verbs and articles are also expected in individuals with HD (Jensen, Chenery, & Copland, 2006).

Impacts on Swallowing Perspective (Dysphagia):

The impacts of HD on the swallowing perspective can be seen at the first level of HD during which little and irrelevant swallowing difficulties are notable. In the second level, the swallowing difficulties become more notable in an individual with HD. At the third level, the swallowing difficulties contribute significantly to the health state of the person.

There are many problems with the swallowing mechanism in individuals with HD which may affect the different phases of swallowing and therefore result in dysphagia. For Instance:

At the *oral phases* of swallowing, an individual with HD may present with weakness in the labial area, tongue instability and difficulties in bolus manipulation. At the *laryngeal phases* of swallowing, an individual with HD may present with delay in laryngeal elevation and epiglottic cartilage (Reyes, Cruickshank, Thompson, Ziman, & Nosaka, 2014).

At the *esophageal phases* of swallowing, an individual with HD may present with abnormally viscous bolus transit and esophageal inflammation associated with gastroesophageal reflux. Swallowing difficulties are considered among the most problems which may affect the lives of people with HD. Some studies have noted that 86% of individuals with HD die due to aspiration pneumonia (Heemskerk & Roos, 2012).

Treatment Plan:

Speech, language and swallowing disorders are usually diagnosed and evaluated by the Speech-Language Pathologist. The evaluation and treatment seek to contribute the communicative wellbeing and improve the quality of life.

In general, treatment of speech and language disorders is a complex process in all neurodegenerative diseases, including HD. The complexity of treatment usually lies in coordinated interdisciplinary work with different health professionals who work on an appropriate treatment plan (Veenhuizen & Tibben, 2009).

From the speech perspective, the intervention program usually involves respiratory retraining to increase the breathing efficiency during speech (Zinzi et al., 2007), which will improve voice aspects. In the same way, orofacial musculature retraining may preserve mobility in both speech and swallowing aspects as well as facial expression.

Huntington disease (HD) affects individuals in different ways and each individual is a unique case in terms of treatment (Hamilton et al., 2012). However, the treatment of speech and communication in HD disorder can be summarized into three simple levels:

1st Level: breathing exercises and muscle relaxation.

2nd Level: family counselling or caregivers counselling regarding adjustments of communication in daily life activities.

3rd Level: implementation of AAC strategies and systems to increase communicative effectiveness (Ferm, Sahlin, Sundin, & Hartelius, 2010), and increase in the individual's participation in social activities (Rusz et al., 2013).

From the swallowing perspective, individuals with HD usually follow essential strategies prescribed by a speech therapist in order to facilitate and guarantee effective safe swallowing.

There are several approaches and techniques which help swallowing problems in individuals with HD, including:

Compensatory Manoeuvres

Approach: this approach usually involves re-education of swallowing and modification of eating behaviors. For example, placing solid foods on molars to ensure that they are crushed well without remaining residuals that could exacerbate difficulties deglutition (Heemskerk & Roos, 2012).

Food Consistency Modifications

Approach: this approach is a fundamental approach as there is a close relationship between food viscosity and aspiration probability (Clavé, Arreola, Romea, Medina, Palomera, & Serra-Prat, 2008).

Sensory Stimulation Approach: this approach usually uses cold and acidic substances in order to stimulate swallowing in oral phases (improves the onset) and pharyngeal (reduces the delay). Also, helps in decreasing involuntary oropharyngeal movements and the frequency of aspiration of the food bolus (Pelletier & Lawless, 2003).

Postural Compensatory Approach: this approach aims to minimize involuntary movements and increase safety food intake (Kagel & Leopold, 1992) through the proper position of the extremities, such as neck and chin. Chin Tuck maneuver is an example of this approach (Nance, 2012).

Conclusion:

Huntington's disease (HD) is a neurodegenerative disorder that directly affects speech and language mechanisms. These may include breathing difficulties, voice disorders, articulation disorders, resonance disorders, fluency disorders and difficulties in receptive and expressive language. The implications of HD can also involve swallowing problems of individuals. Thus, a speech language therapist can help identify and detect these difficulties and assist in improving the quality of life and the communicative wellbeing of individuals with HD.

In Saudi Arabia, most speech language pathologists work in main hospitals located in the three major cities (Riyadh, Jeddah and Eastern region). Huntington Disease cases may not receive minimal care or diagnosis or treatment plan for speech, language and swallowing aspects. As mentioned before, the purpose of this article was to highlight the difficulties and implications that face individuals with HD in general. Reports and studies about HD in Saudi Arabia are very limited, thus, this article may be considered as an initiative to shed light on the implications of Huntington's disease on speech, language and swallowing in Saudi Arabia.

Future research and studies are needed to enhance scientific research regarding HD in Saudi Arabia.

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SSSPA Values

To provide the latest evidence based information to professionals in order to improve their knowledge base and clinical skills and to further develop their expertise. Support and participate in research which would facilitate and enhance clinical practice. Start initiatives with governmental and private organizations so that their policies would fulfil professional needs and would enhance their capabilities to deliver high quality services to individuals with hearing, balance, speech and swallowing disorders. Promote the improvement of quality of life for individuals with hearing, balance and speech disorders and empower them to participate in all aspects of the community (social, financial and cultural). Provide consultations to scientific and occupational organizations as well as to individuals in the community. Outreach to all national and international scientific health associations/societies in order to support the profession, SSSPA members and the community. Enhance the efficiency within SSSPA and encourage all members of SSSPA to effectively participate in all its events. Continue to function and collaborate with all partners with transparency.

Upcoming Events

- ◆ 11th International Saudi Otorhinolaryngology conference to be held in February 14- 16, 2019 at Le Meridien Hotel, AlKhobar, Saudi Arabia.
- ◆ The first Advanced Bionics Audiology Course in Saudi Arabia, 25-27/02/19 at King Abdulaziz University Hospital.
- ◆ The SSSPA 6th Scientific Club Meeting .
- ◆ The Second Let Us Debate Competition.

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