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Deaf with Waardenberg Syndrome Type I: Jordanian Case Study

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ABSTRACT

This study aimed to identify deaf people with deafness of type 1 Waardenburg syndrome. To achieve the study objectives, use the qualitative method for a single case study. The sample included (11) deaf students from all deaf schools in Jordan, whose ages ranged from (8-19), and (Wechsler Intelligence Scale, WISC-IV) in sign language was applied to them. The results showed that they had symptoms in terms of sensorineural hearing loss .And the divergence of the eyes from the inner corner. On the front of the head, and on the eyelashes or on the eyebrows. With variations in the pigment of the iris, their eyes are blue, brown or black. Also, the root of the nose is wide. And that the emphasis during communication with sign language is on the hands of the speaker and puts a distance during communication to clarify the vision of the signal. Further studies and research on the level of intelligence of deaf people with type 1 syndrome, visual perception studies in deaf people with type I Waardenburg syndrome, and the inclusion of this syndrome as an important part of the categories of hearing impairment are recommended for the study.

Keywords
Deaf, Wardenberg syndrome, Hearing impairment syndromes, Sign language, WISC-4.

I. Introduction

This specialized research for Waardenburg syndrome is the first type for people with hearing disability from rare Arab research (according to the researcher's knowledge) and this in turn confirms that the category of hearing disability is a heterogeneous group, according to the degree of auditory loss, and the syndrome is considered a non-infectious disease, and it cannot be treated with drugs And it cannot happen due to lifestyle or growth factors, as many books specializing with hearing impairment and theoretical literature that are almost identical have not indicated and they were not educationally discovered, which led to the emergence of a new pediatric syndrome known as Waardenburg syndrome within the class. Deaf, if any, the information provided is almost insufficient to know the truth of this syndrome. Therefore, the measurement and diagnosis of Waardenburg syndrome for people with hearing disability is one of the issues whose folds have aspects of great difficulty, whether it is medical (for the cost of examining the gene that causes the syndrome) in general or psychometric (because there are no specialized measures for people with auditory disability of Waardenburg syndrome whether it is for mental ability, perception, Sign Language) or pedagogical standards that are appropriate to their ability to use sign language. Hence the current study came to provide an explanation of the characteristics of deaf people with Type I syndrome by knowing the symptoms and sign language for them.

In (1947), the disease was discovered by the Dutch ophthalmologist Dr. Petrus Johannes Waardenburg, where he observed that the color of the eyes of some of those who had designs differed from each other. Although the prevalence of this disease reaches one case per (20000-40000) new born children, it affects approximately (3%) of the total of those who have been designed since birth and their characteristics were as follows:

- Congenital hearing loss.
- The presence of a side shift to the inner corner of the eyes.
- Variation in skin pigmentation (albinism).
- White premature graying of hair at the front of the head.
- The eyebrows are bound together (excess eyebrows sticking together in the midline).
- Blue eyes and pigment color differences in the iris (Heterochromia Iridium).
- The root of the nose is wide. (Waardenburg, 1951).

Until this syndrome became known as Waardenburg Syndrome Type 1, WS1. Then Arias (1971) defined Waardenburg syndrome of the second type (WS2). As individuals of the second type (WS2) have all the characteristics of the first type (WS1) except for the angle of the eye. (Pantke, Cohen, 1971). The classifications of this syndrome have also evolved with different symptoms, and therefore we can differentiate between those subtypes of Waardenburg syndrome and

work on knowing ways of communication that are appropriate to the type and severity of the problem. Waardenburg syndrome, WS syndrome is a rare and unusual disease characterized by sensory nerve deafness, pigmented deformities as well as defects of auditory nerves. With additional symptoms, Waardenburg syndrome is classified into four types (WS1, WS2, WS3, and WS4), which are called:

1- Waardenburg Syndrome Type 1, WS1.
2- Waardenburg Syndrome Type 2, WS2.
3- Waardenburg syndrome of the third type, also called Klein-Waardenburg syndrome, (Waardenburg Syndrome Type 3, WS2).
4- Type 4 syndrome, also called Waardenburg-Shah, Waardenburg Syndrome Type 4, WS4.

Waardenburg syndrome is considered to be a heterogeneous syndrome. Doctors have identified four types with different symptoms. Individuals with this syndrome have symptoms similar to each type, but type I and II are the most common for this syndrome:

1- Waardenburg syndrome type 1:

This syndrome is characterized by the fact that the individual has a large distance between his eyes. Where (20%) of them suffer from hearing loss, especially the inner ear. They have a difference in the color of the iris or the eyes together, and the appearance of white hair on his head, whether it is at the front of the head or a dispenser (Hart, Miriyala, 2017; Abu Drei, 2020).

2- Waardenburg syndrome types two:

Hearing loss is more common in this type compared to type I, and their hearing loss is (50%). Symptoms are similar to type I, including changes in hair, skin, and eyes. This type is distinguished from the first type mainly because there is not much distance between the eyes (Jalilian et al., 2018; Abu Drei, 2020).

3- Waardenburg syndrome type III:

measuring the distances between the inner corners of the eye and the outer corners of the eye, by means of a (W) indicator to check whether or not the angle of the eye (Dystopia canthorum) is present. Which is the result that it may suggest that we have Waardenburg syndrome type I is as follows?
1- The distance to the inner eye angle (a).

The third type is similar to the first and second types, and often results in loss of hearing and a change in the pigment of the iris of the eyes. People who own this model usually have a great distance between their eyes and their wide nose. The distinguishing feature of this type that distinguishes it from the first and the second type is the presence of problems with the upper limbs, as those with the third type have problems and deformities in the arm or shoulders or deformities of the joints. Some also suffer from mental disabilities or the rabbit's lip accompanying this type. The third type is also called Klin-Waardenburg syndrome (Klein, Opitz, 1983).

4- Waardenburg syndrome, type IV:

Type IV causes changes in the pigment of the iris and may also lead to hearing loss. People with this type of disease have a colon disorder, which causes severe constipation. (Shah et al., 1981).

Characteristics of deaf people with Waardenburg syndrome

Waardenburg (1951) notes 6 main characteristics related to Waardenburg syndrome types: Wide nasal root, partial or total pigmentation differences in the iris of the eye, from birth, brow attachment, and white hair to the front of the head (Tagra et al., 2006). In most individuals, Waardenburg syndrome (WS1) is diagnosed by performing a physical examination to search for clinical criteria that include:

- Loss of sensory hearing.
- Pigment changes in hair and eyes. (Shields et al., 2013)

This syndrome can be recognized by calculating the Waardenburg index (W), which is the necessary measurement for the calculation of the W (mm) index. In the case of suspicion of Waardenburg syndrome, an appropriate diagnostic evaluation may include the use of a caliper (used to measure thickness or diameter), This syndrome can be diagnosed by
2- The distance between the two pupils (b).
3- The outer distance of the angle of the eye (c).
It is through the following formula:

$$X = (2a - (0.2119c + 3.909)) / c$$
$$Y = (2a - (0.2479b + 3.909)) / b$$
$$W = X + Y + a / b$$

Mathematical results are an eye angle greater than (W <1.95 index).
Through this equation, the diagnosis of Waardenburg syndrome can be measured from birth or early childhood through comprehensive clinical assessment and determination of results and their physical characteristics. (Farrer et al., 1992). He also indicated (Pingault et al, 2010) that Waardenburg syndrome is the leading cause of symptomatic sensorineural hearing loss (SNHL) and is a disease that causes visual disturbance, eyebrows, iris divergence, and white hair.

The US Department of Health and Human Services, National Institute of Deafness and Other Communication Disorders (NIDCD, 2013) defines Waardenburg syndrome as a rare genetic disorder characterized by hearing and pigmentation mutations. Harlor & Bower (2009) confirms that Waardenburg syndrome is a high sign of hearing loss. Even if newborns are examined, and the American Academy of Pediatrics recommends referring at least one diagnostic evaluation, These children are also evaluated no later than every (3) months of age. Hearing loss requires appropriate referral through (otolaryngology, audiology, genetics) and intervention no later than (6) months of age.

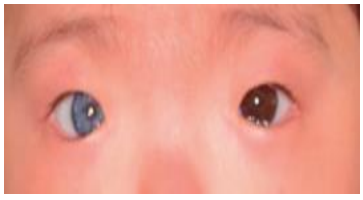
Waardenburg syndrome usually has distinct changes in hair and skin, as well as sensory nerve hearing loss. In this minor variation, symptoms may be hidden and may not be diagnosed until either a family member is diagnosed or all family members are examined. (Egbalian, 2008).

Deaf people with this syndrome have eye changes such as each eye in a different color from the other, or hair or skin, which can be seen easily. The degree of hearing loss is not always constant but varies from simple to severe hearing impairment and may be mono or bilateral. (Toriello et al., 2004). In (2009) (Adameyko et al., 2009) presented three sets of evidence on a large number of pigment cells and how they affect the skin. Dermatological analysis (congenital birthmarks) has led to the emergence of a new theory proposing the existence of a new group of primary cells derived from melanocytes originating in the mesoderm, which arises due to centrifugal migration.

The melanocytes are almost complex, and are responsible for the color difference of the skin, eye (iris), skin, and cochlea (Gorlin et al., 2001). Also (Steel et al., 1989) confirms that Waardenburg syndrome affects physical features due to the absence of melanocytes, especially in the inner ear (cochlea) through the vascular layer, that the hearing will be affected. Also (Hager et al., 2010) indicates that Waardenburg syndrome is a rare disease characterized by loss of sensory nervous system and chromosomal abnormalities of the iris, skin and hair due to mutations in the gene (PAX3).

Each (Laababsi, Allouane, Beghdad, Rouadi, Abada, Roubal, Mahtar, 2018) conducted a one-case study entitled "Waardenburg Syndrome: A Rare Cause of Auditory Sensory Impairment in Newborns" aimed at identifying and considering Waardenburg Syndrome as a rare disease characterized by deafness and associated with the presence of chromosomal abnormalities and defects in nerve tissues derived from the nerve. It is responsible for (2%) of severe congenital hearing loss. The sample included a 4-year-old child from Morocco, who is a single child in the family (not from consanguineous marriage), and the results of the clinical examination showed the following:

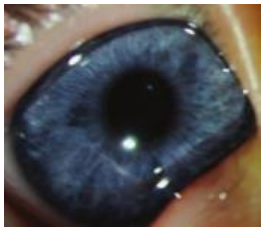
- He has suffered from a loss of hearing in both ears since childhood (100 dB).
- Delayed speech development.
- The child has the characteristics of Waardenburg Syndrome Type 1.
- He had a cochlear implant.
- The eyes are blue (sapphire).
- There is no infection of the eardrum.
- There are no other abnormalities of the musculoskeletal system or of Hirschsprung's disease.
- Visual acuity (10/10) in both eyes.



The child's psychological state is good.

And (Yoshida et al., 2016) conducted a study entitled "A New (PAX3) Mutation in a Japanese Child with Type 1 Waardenburg Syndrome" aimed at identifying a new gene for Type 1 Waardenburg Syndrome, which is a rare autosomal inherited disorder characterized by a deficiency of hair pigmentation and abnormal iris pigmentation. And congenital hearing loss. Type 1 is caused by mutations in the (PAX3) gene. We identified a novel (PAX3) mutation (c.1107 C> G, p.Ser369Arg) in a Japanese patient with type 1 Waardenburg syndrome. The results showed after conducting genetic and clinical tests:

- Abnormal iris pigmentation.
- Congenital hearing loss in the right ear.
- The eyebrows are joined together (unibrow).



Incidence of deaf people with Waardenburg syndrome indicate that the incidence of Waardenburg syndrome has been reported in societies, indicating that the clinical characteristics of two categories: one of European origin and the other of South Asia. Where the report indicated that Waardenburg syndrome is between (2 - 5%) of cases of congenital deafness. It was first described in the northern European regions, particularly for those with white skin. (Nayak, Isaacson, 2003)

Oiso et al. (2013) considers the Waardenburg syndrome a hearing disorder that affects between (2 -3%) congenital deafness. Estimated cases around the world are (2 to 3 cases per 100,000) of society, and equally affect both sexes.

The American National Institute of Medicine (Vichare, Bhargava, 2013) indicates that not everyone with Waardenburg has a hearing loss. It usually affects only hearing impaired (60%) of those diagnosed with the syndrome.

And (Black, Pesznecker, Allen et al., 2001) conducted a study entitled "Vestibular System Pattern of Waardenburg Syndrome" aimed at identifying the vestibular anomalies associated with Waardenburg syndrome. The sample of the study included (22) white-haired adults with a clinical diagnosis of Waardenburg Syndrome as (10) from the first type and (12) from the second type. Where the auditory vestibular function was diagnosed. Results showed vertigo (dizziness) and imbalance. Also, test results including (caloric, vestibular automatic rotation, and semi-random rotation) were abnormal (77%), and vestibular functional test results were abnormal (57%), but there were no specific patterns of abnormalities. Whereas, (6) of the individuals have a sensorineural hearing loss. And (3) individuals suffering from severe hearing loss, and the results also showed that the cerebral hearing response was normal. And that individual with Waardenburg syndrome may experience vestibular symptoms in the first place without hearing loss. Electrocochleography and vestibular function tests are the most sensitive measures of the presence of ear abnormalities for these individuals.

Song et al. (2016) conducted a study entitled "Hearing loss in Waardenburg syndrome: a literature review" aimed at identifying auditory and genetic problems of the syndrome. Calculate the prevalence rates of auditory loss associated with different types and genes of Waardenburg syndrome. A sample included (73) articles, and (417) patients were described. The results showed that:

- Auditory loss was (71%) and was significantly associated with Waardenburg syndrome.

Mental ability in deaf people with Waardenburg syndrome.When talking about the relationship between Waardenburg syndrome and mental disability, there is a difference about the mental ability of Deaf people with Waardenburg syndrome.

And (Chen, Harold, 2006) indicates that Intellectual Disability and developmental abnormalities were evident in nearly all patients.

And also (de Saxe, 1984) found that families affected by the syndrome found delayed or impaired academic performance (achievement) that required special education for (9) persons associated with the Waardenburg syndrome.

The report (Kawabata et al., 1987) refers to a case study of a (13) year-old child who had a set of symptoms for Waardenburg syndrome, including:

- Mental retardation.
- Significant movement delay.
- Severe disturbance in gait.
- Dysfunction of muscle tension.
- Muscle sclerosis and peripheral neuropathy.
- Another report (Pasterls, Trask, Sheldon, Gorskl, 1993) of a case study of Waardenburg syndrome suggests that there are symptoms including:
- Small size of the head.
- Having a mental disability.
- Severe physical disorders and abnormalities.

Also (Kiani, 2007) asserts that deaf individuals (prior to language acquisition) with the Waardenburg Syndrome have not reported this relationship in the theoretical literature. We believe that there is a striking similarity to these two people, which provides us with the possibility of apparent behavioral patterns by describing the cases with the presence of:

- Fragile (X) syndrome.
- German measles.
- Herpes simplex encephalitis.

Although there are no Arab studies (according to the researcher's knowledge) that confirm the understanding and characteristics of deaf people with Waardenburg syndrome of the first type, those cases were discovered when (Abu Drei, 2017) conducted an extensive study entitled "Standardization A Jordanian Version of The Wechsler Intelligence Scale " for Children -Fourth Edition Adapted for Deaf Via Sign Language From (6 - 16.11) years " This study aimed to legalize the Jordanian image of the (wisc-4) at school stage for the age group (6 - 16.11) years to measure intelligence in sign language for the deaf. In order to achieve the objectives of the study, the wisc-4 IQs were converted after indications of their validity, reliability, and criteria of the wisc-4 scale in the ordinary and deaf samples were converted into sign language. The sample of the study included the deaf and ordinary (831), and indications of the validity of the Jordanian image from the (wisc-4) scale in the sign language of the deaf were represented in the honesty of the content, discriminatory honesty, sincerity of the building, and associative honesty, as well as indications of consistency The scale was the use of Alpha Cronbach, the method of return, and the residents' agreement.

It was also reached to the criteria of the scale represented by converting the raw grades to standard degrees and then to an IQ. The results of the study showed:

- There were statistically significant differences (α = 0.05) in performance on the sub-tests of the Jordanian image of the scale (wisc-4) of children's intelligence in the school stage due to a variable in the ordinary and deaf category and the gender variable in the deaf group.
- There were statistically significant differences (α = 0.05) in performance on the sub-tests of the Jordanian image of the scale (wisc-4) for deaf children in the language of sign at school due to the change in the degree of hearing disability (simple - medium - severe - cochlear - simple + cochlear) for the category Age (6-16.11) years.
- There are statistically significant differences in performance on the subtests of the Jordanian image of the scale (wisc-4) of children in school due to the age variable for the deaf in the sign language in favor of the age group (13.00-13.12).

On this syndrome in the deaf, (Drei, 2020) conducted a study entitled "Deaf People with Waardenberg Syndrome Type II: Jordanian Case Study" which aimed to identify deaf people with deafness with Waardenburg Syndrome type II. In order to achieve the objectives of the study, the qualitative method was used for the one case study. The sample included (2) deaf students from all deaf schools in Jordan, who are between the ages of (9-10), and (Wechsler Intelligence Scale ,WISC-IV) was applied in the language of Performance Scales and Audiometry. The results of the study showed the presence of symptoms in Deaf people with Waardenberg syndrome type II in terms of: auditory sensory loss. The presence of a side shift to the inner corner of the eyes. Premature graying of hair on the front of the head in white and some on the eyelashes or on the eyebrow. The eyebrows are connected to each other. Dark blue or brown eyes with black and pigment color differences in the iris. The root of the nose is wide and the presence of the rabbit's lip. The results of the study also showed that there is an intense focus during communication in the sign language at the hands of the speaker due to the fact that the color contrast of the eyes led to a dispersion of the focus in the sign language. They also have a sense of the direction of light, which may adversely affect the process of visual communication of sign language. The results of the study also showed that the average hearing impairment

was the highest category. The study recommends further studies and research related to the level of intelligence of deaf people with Type II syndrome, as well as studies related to visual perception in Deaf people with type 2 Waardenburg syndromes and the inclusion of this syndrome as an important part of the categories of hearing impairment.

Research Questions:

The study answers the following questions:

1- What is the concept of Deaf people with Waardenburg syndrome type I on a Jordanian sample?

2- What are the characteristics of Deaf people with Waardenburg syndrome type I on a Jordanian sample?

3- What is the level of hearing impairment for Deaf people with Waardenburg syndrome type I on a Jordanian sample?

Study Significant:

Knowledge of Deaf people with Waardenberg syndrome type I was provided on a Jordanian sample.

First: theoretical importance:

1- Arouse interest in the knowledge of Waardenburg syndrome for people with hearing impairment.

2- Knowing the relationship between the degree of auditory loss and Waardenburg syndrome of the first type.

3- The scarcity of Jordanian studies that deal with Deaf people with Type I syndrome.

4- Enriching the Arabic library with a category related to hearing disability.

Second: The practical importance:

1- Determining the characteristics of Deaf people with Waardenburg syndrome type I.

2- Helping researchers benefit from the results of this study in dealing and communicating with Deaf people with Waardenburg syndrome and determining their mental ability.

Study limitations:

This study has some limitations:

1- The scarcity of Jordanian studies that deal with deaf people with Waardenburg syndrome Type I.

2- The scarcity of studies dealing with sign language with Deaf people with Waardenburg syndrome Type I.

Definitions of Terms:

1- Deaf : They are individuals who use sign language, alphabet, lip language, and the total way to communicate with each other, whether with or without the use of auditory aids, and their hearing aids range from (25 - 90) decibels.

2- Waardenburg syndrome, the first type : it is a group of rare features that appear on the individual that distinguishes them from others, which include the expansion of the root of the nose, which causes expansion of the eyes, and the appearance of a difference in the iris color of both eyes, which makes the shape of the face unusual and the presence of other changes in their appearance that are White hair (gray hair) in early childhood until before (30) years, as well as auditory loss, and the presence of some developmental problems.

Methodology: Method and Procedures

Method of study

A qualitative approach was used to suit the purposes of the current study, which is based on the evidence of the interview and observation, in addition to relying on the (coding) test from the Jordanian image of the WISC-4 scale for deaf intelligence in the sign language to determine the level of visual problems among study members.

The sample of study

Qualitative studies depend on a limited number of respondents, whereby the study members were identified through the researcher's follow-up of deaf cases from a community of (947) deaf students, and the researcher working in the field of hearing disability diagnosis Their number (n = 11) is singular within the age group (8-19 years), who were chosen intentionally from the total community of the deaf, and registered in schools of education.

Repeats and percentages were used to describe the study sample members

Table 1. Distribution of study sample individuals according to age

percentage	Repetition	Age
9.09	1	8
27.3	3	9
9.09	1	10
18.2	2	11
9.09	1	12
18.2	2	16
9.09	1	19
100.0	11	Total

Table 2. Distribution of study sample individuals according to gender

percentage	Repetition	gender
54.54	6	Male
45.45	5	female
100.0	11	Total

The study tool

1- Case study questionnaire (syndrome symptoms): A list of symptoms for this syndrome was prepared through theoretical framework and previous studies of the syndrome, which include:

- Congenital hearing loss.

Research procedures:

To achieve the aims of the study, the following measures were taken:

1- Syndrome symptoms were identified for deaf students.

2- The results (n = 2) of Deaf people with Waardenburg syndrome type I were extracted.

3- Deaf students with Waardenburg syndrome type I were divided into classes according to the variable degree of hearing impairment (simple, moderate and severe).

4- Symptoms of the syndrome were depicted on (10) deaf people with Waardenburg syndrome type I, out of (11) for rejection of the latter from the imaging process.

5- The performance section of the (WISC-4) scale was applied to the deaf.

The Study Results:

Study questions will be answered according to their sequence:

- The presence of a side shift to the inner corner of the eyes.
- Variation in skin pigmentation.
- White gray hair on the front of the head.
- The eyebrows are bound together.
- Blue eyes or pigment color differences in the iris.
- The root of the nose is wide.

2- Auditory examination.

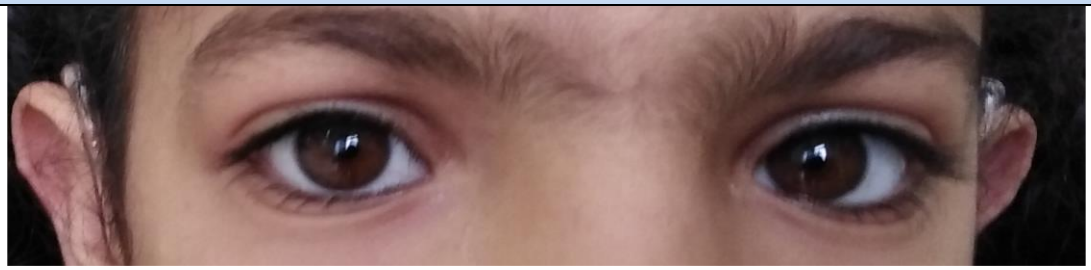
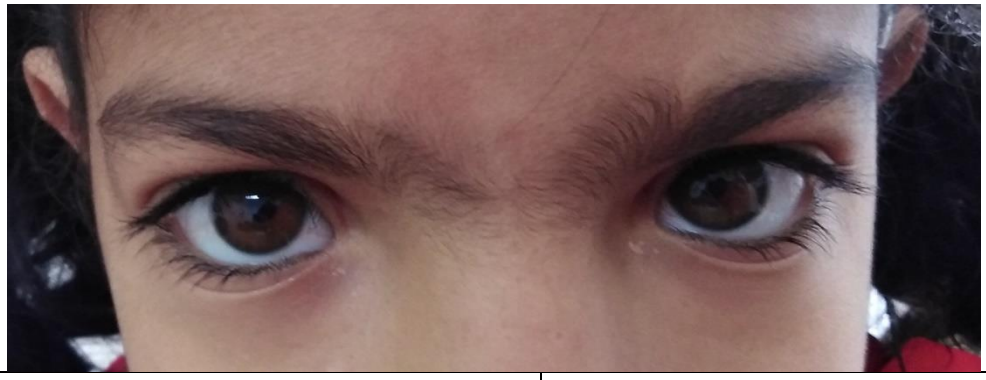
3- (WISC-4) Scale: Performance Section (Coding Test).

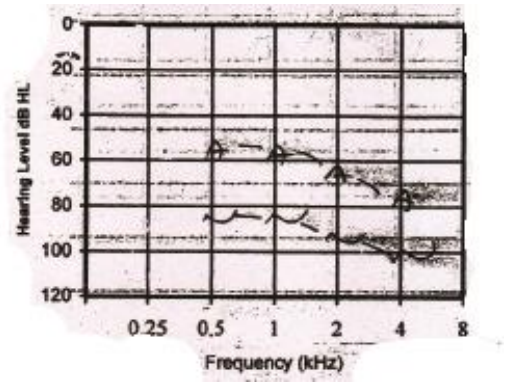
Results for the first question: What is the concept of deaf people with Waardenburg syndrome type I on a Jordanian sample?

The researcher defined the concept of deaf people with Waardenburg syndrome type I through a set of symptoms that have been studied on deaf in terms of:

- Congenital sensory hearing loss.
- The presence of a side shift to the inner angle of the eyes (through the calculation formula previously mentioned).
- Variation in skin pigmentation.
- Premature graying of hair on the front of the head in white and some on the eyelash or on the eyebrow.
- The eyebrows are bound together (excess eyebrows sticking together in the midline).
- Blue eyes or dark brown with black and pigment color variations in the iris.
- The root of the nose is wide.

And through the following cases we review the following:

CASE (1): Type I (WS1), Age: 8 years, Gender: female	
	
	
Auditory Test	**Symptoms**

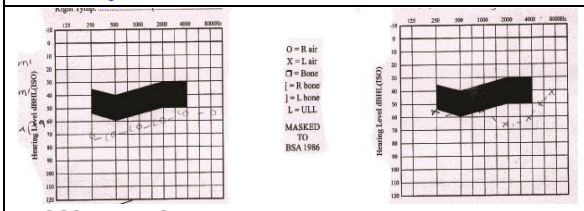


Severe hearing loss

- The bridge of the nose is very wide.
-eyes spaced.
- Eyebrows bond together in the middle.
- Pigmentation of some parts of the eye (brown with black).

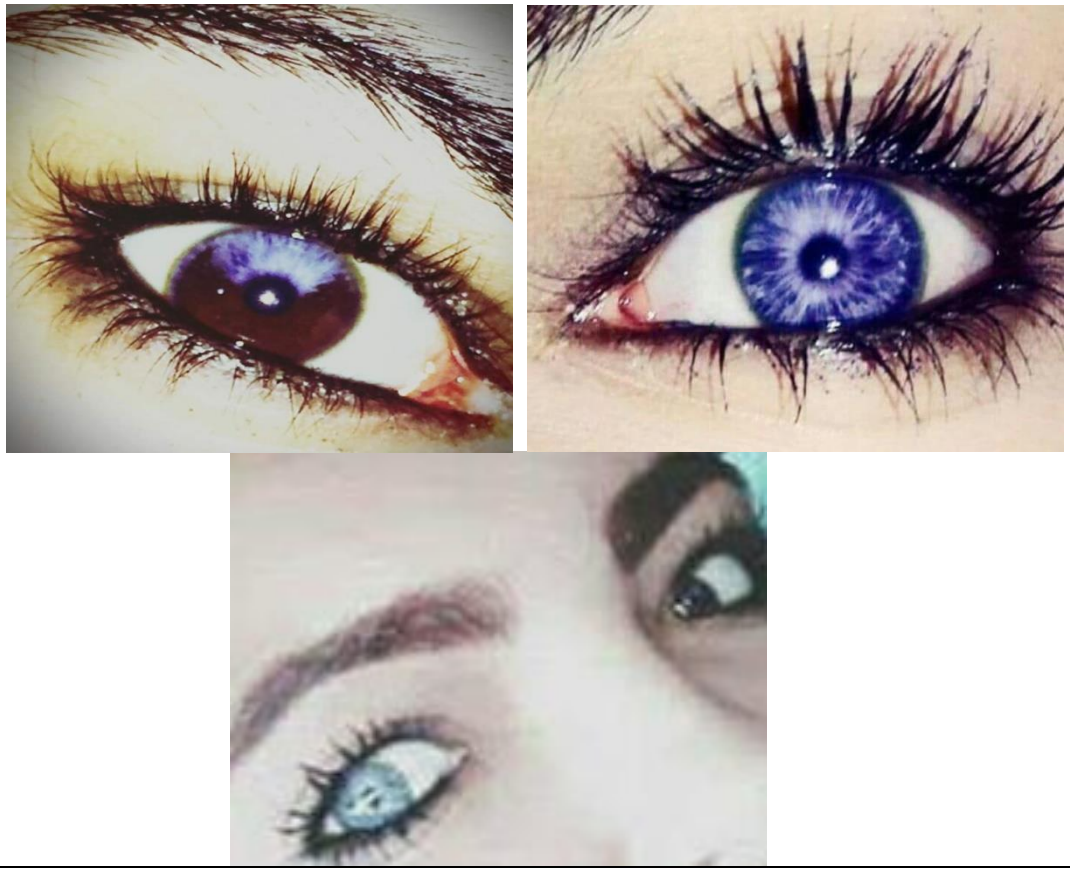
CASE (2): Type I (WS1), Age: 11 years, Gender: female

There is no picture of the case

Auditory Test	**Symptoms**
 mild hearing loss	- The bridge of the nose is very wide. - Eyebrows bond together in the middle. - A white tuft at the front of the head.

CASE (3): Type I (WS1), Age: 16 years, Gender: Female

Left eye Right eye



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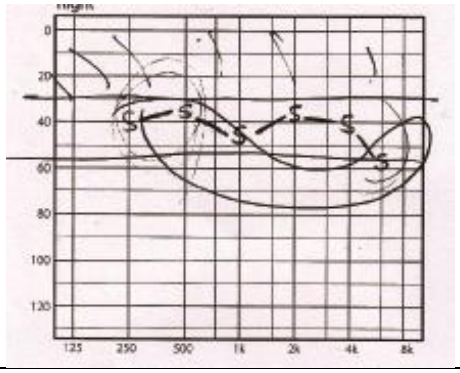
Auditory Test	**Symptoms**
mild hearing loss 	- The bridge of the nose is wide. -The eyebrows were joined together in the middle, but was removed due to its thickening. - White tuft on the side of the head in the middle. -Patches on the skin in the arm and shoulder area.

CASE (4): Type I (WS1), Age: 16 years, Gender: Male

Left eye

Light blue with a small black spot

Right eye

Half of it is dark blue, the other half is black, and the shape of the pupils is different from the right.

Auditory Test	**Symptoms**
Severe hearing loss	- The bridge of the nose is very wide. -The eyebrows are joined together in the middle.

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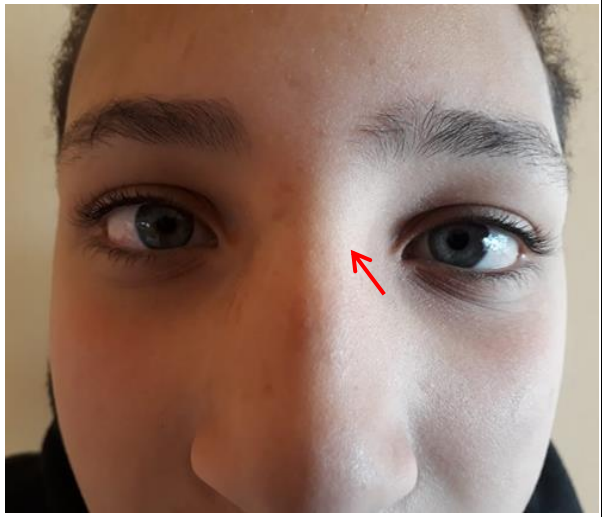
CASE (5): Type I (WS1), Age: 11 years, Gender: Male

Left eye

The eye is different from the right

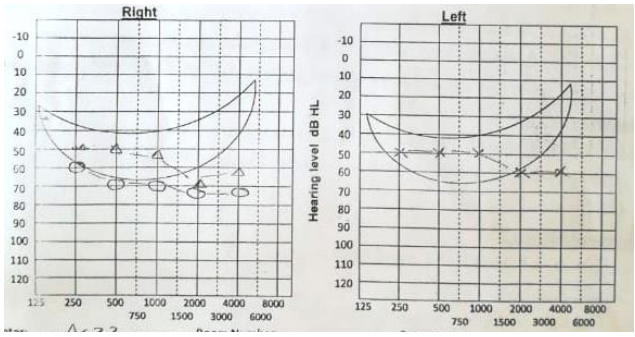
light blue

Right eye



Light blue with a dark blue spot, and the pupil has a different shape than the left eye.

Auditory Test



mild hearing loss

Symptoms

- The bridge of the nose is very wide.

CASE (6): Type I (WS1), Age: 10 years, Gender: Male

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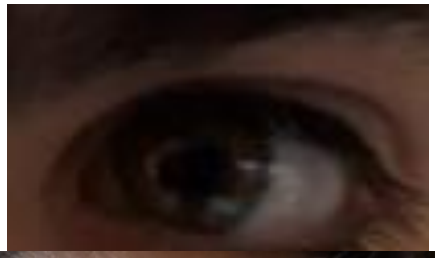
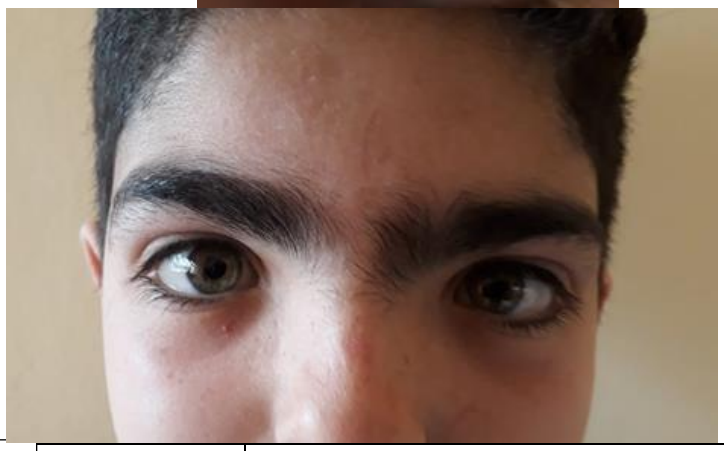
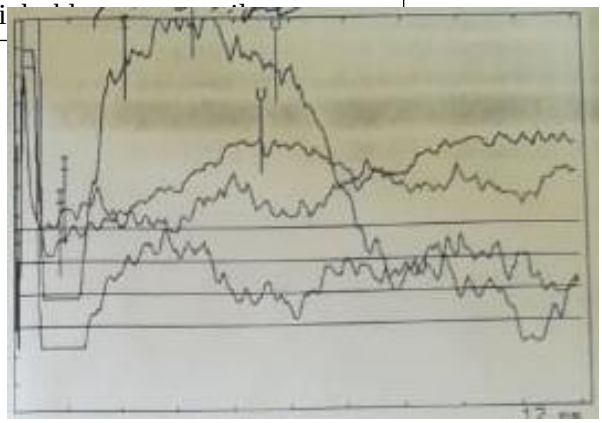
Left eye	Right eye
	 
Light green with dark spots and a li…  mild hearing loss	**Symptoms** - The bridge of the nose is very wide. - The eyebrows are joined together in the middle.

CASE (7): Type I (WS1), Age: 9 years, Gender: Male

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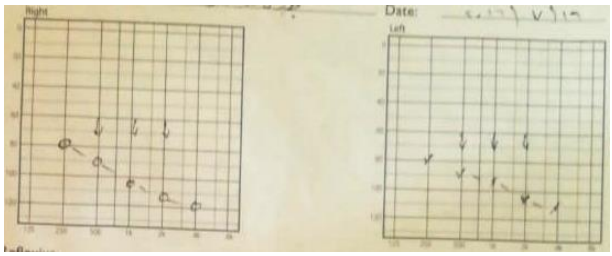
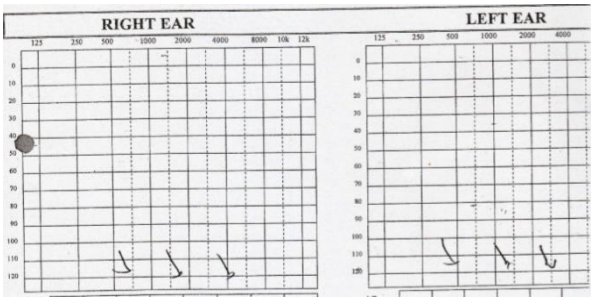
Left eye	Right eye
	
The slightest degree of blackness	The color is dark black

Auditory Test	Symptoms
 Moderate hearing loss	- The bridge of the nose is very wide. - The eyes are wide apart. The eyebrows are joined together in the middle.

CASE (8): Type I (WS1), Age: 12 years, Gender: Female

Left eye	Right eye
	
The lowest degree of the left eye and the presence of drawings of the eye different from the right.	

Auditory Test	Symptoms



Moderate hearing loss

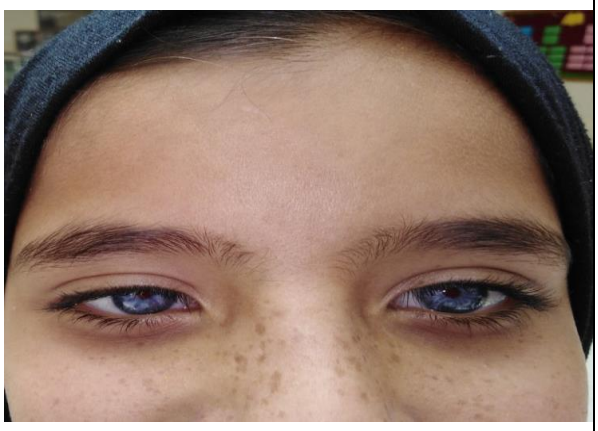
- The bridge of the nose is wide.
- The eyes are wide apart.
- The eyebrows are slightly intertwined.
- White locks at the front of the head (difficult to photograph because of the veil).

CASE (9): Type I (WS1), Age: 9 years, Gender: Female

Left eye

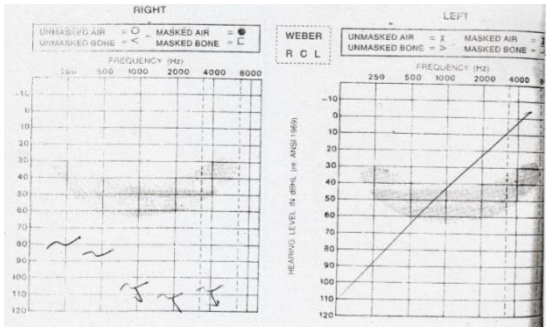
Black color interferes with dark blue.

Right eye



Minimal black in the lower parts. And the presence of blue as a mosaic.

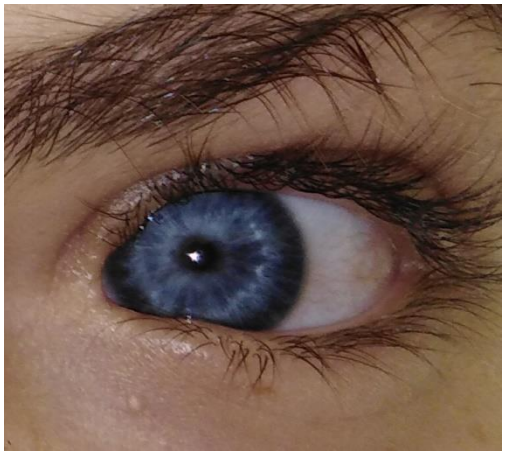
Auditory Test	**Symptoms**

 Moderate hearing loss	- The bridge of the nose is very wide. - The eyes are wide apart. - The eyebrows are joined together. - Tufts of white hair at the front of the head.

CASE (10): Type I (WS1), Age: 9 years, Gender: Female

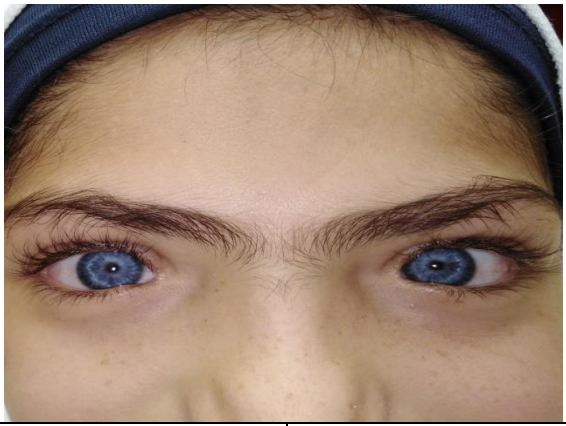
Left eye

Right eye

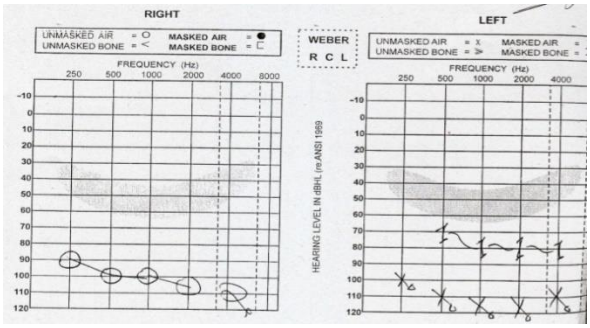


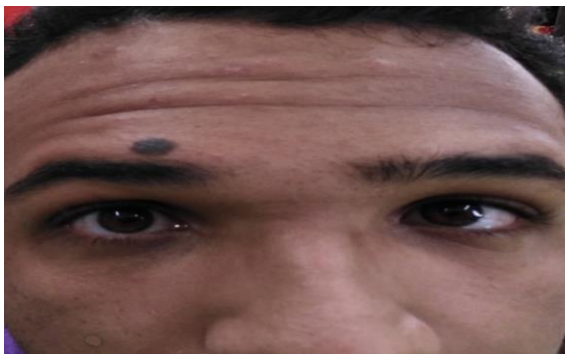
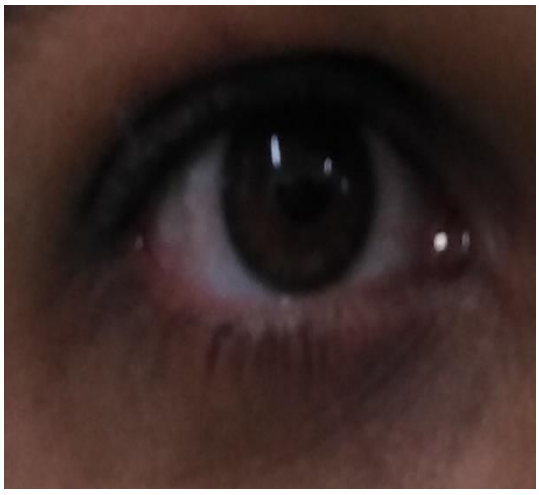
A degree lower than the left.

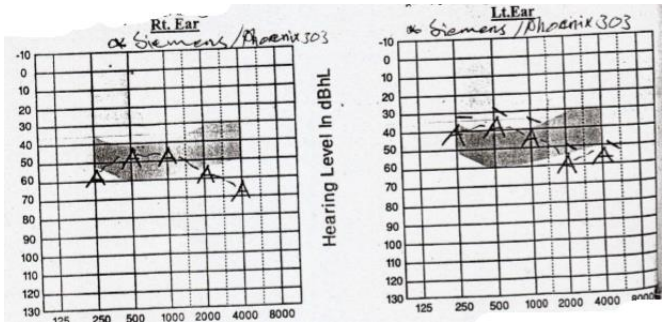
Dark blue



Auditory Test	**Symptoms**

 Moderate hearing loss	- The bridge of the nose is very wide. - The eyes are wide apart. - The eyebrows are joined together in the middle and dense. - The presence of some white tufts on parts of the front of the head.

CASE (11): Type I (WS1), Age: 19 years, Gender: Male	
Left eye  A degree lower than the left	Right eye  Dark brown and black pigmentation on the sides of the eye.
	
Auditory Test	**Symptoms**

 Moderate hearing loss	- The bridge of the nose is very wide. - The eyes are wide apart.

Results related to the second question: What are the characteristics of Deaf people with Waardenburg syndrome type I on a Jordanian sample?

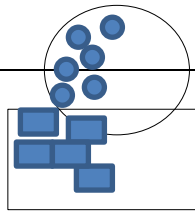
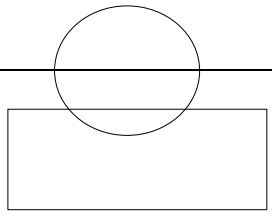
Deaf people with Waardenburg syndrome type I showed some characteristics that deaf their deaf peers from other groups:

First: Characteristics of sign language:

- Intense focus while communicating in sign language at the hands of the speaker due to the fact that heterogeneity of eyes led to a dispersion of focus in sign language.
- Putting a space during sign language communication to clarify the vision of the sign, especially for deaf people with the right eye blue and the left half of it blue and the other half brown.

Second: optical properties:

Deaf people with Waardenburg syndrome and because of the lack of eye pigmentation, they add some shapes to the image, believing it is correct. For example:

Its meaning for the deaf	How to draw it	code
Circle		
rectangle		

- Deaf people with Waardenburg syndrome type I have a sensitivity to light direction, which may adversely affect the process of visual communication of sign language.
- By applying the Wechsler-4 scale, the performance section to some cases of Waardenburg syndrome type 1 through the subtest (coding), note the answer (many of them indicated that there is no defect in the spaces and that they are complete):

Results for the third question: What is the level of auditory impairment for deaf people with Waardenburg syndrome type I on a Jordanian sample?

Frequencies and percentages were extracted to identify the degree of auditory impairment in Deaf people with Waardenburg syndrome type I, and Table (3) shows that:

Table 3. Distribution of study sample individuals according to the degree of hearing impairment

percentage	Repetition	degree of hearing impairment
36.36	4	Mild
45.45	5	Moderate

percentage	Repetition	degree of hearing impairment
36.36	4	Mild
18.18	2	Severe
100.0	11	Total

Discussion:

Discussion of the results of the first question:

Discussion of the first question: What is the concept of deaf people with Waardenburg syndrome type I on a Jordanian sample?

The results of the study showed that there are symptoms in Deaf people with Waardenburg syndrome type I in terms of:

Sensory hearing loss. The presence of a side shift to the inner corner of the eyes. Premature graying of hair on the front of the head in white and some on the eyelashes or on the eyebrow. The eyebrows are bound together. Blue or dark brown eyes with black and pigment color variations in the iris. The root of the nose is wide.

- The results of this study are in agreement with the study (Laababsi et al, 2018), with the presence of distinct characteristics of the Waardenburg syndrome type I, represented by blue eyes (sapphire).
- The results of this study are in agreement with the study (Yoshida et al, 2016), in the presence of abnormal iris pigmentation, congenital hearing loss in the right ear, and eyebrow attachment together.

The researcher explains: Because many researchers have not mastered sign language, this has resulted in the failure to discover cases within the category of hearing disability because it is a heterogeneous group, and the issue of syndromes associated with hearing impairment is limited to medicine. This led to a lack of knowledge of the educational and psychological characteristics and mental capacity of this group.

Discussion of the second question: What are the characteristics of Deaf people with Waardenburg syndrome type I on a Jordanian sample?

The results of the study showed that there is a severe focus during communication in the sign language at the hands of the speaker due to the fact that the color contrast of the eyes led to a dispersion of the focus in the sign language. As well as setting a distance during the sign communication to clarify the vision of the signal, especially for the deaf people with the right eye blue and the left half of it is blue and the other half is brown. They also have a sense of the direction of light, which may adversely affect the process of visual communication of sign language.

- The results of this study are largely in agreement with the study (Drei, 2020) in the presence of sensorineural hearing loss. A sideways deviation towards the inner corner of the eyes. Early graying of hair on the front of the head is white, and some on the eyelashes or on the eyebrow. The eyebrows are connected to each other. Dark blue or brown eyes with variations in black and pigmentation in the iris. The root of the nose is wide. And that there is intense focus during sign language contact on the speaker's hand since the contrast in the color of the eyes led to a dispersion of focus in sign language. They also have a sense of the direction of light, which may adversely affect the visual communication process of sign language.
- The results of this study agreed with (Abu Drei, 2017) that the Jordanian image from the (WISC-4) scale was able to identify some visual problems for this syndrome through performance tests of coding test.
- The results of this study are in agreement with both the study (Laababsi et al, 2018) considering that Waardenburg Syndrome is a rare disease characterized by deafness and associated with the presence of chromosomal abnormalities and defects in the nervous tissue derived from the nerve, as well as the presence of delayed speech development in them.

The researcher explains: that the sign language depends on the use of hands in the communication process, which are rapid, accurate and interlinked signal movements between them and therefore if there are visual problems for the deaf it will affect the process of receiving the language being a visual language and may happen here either not seeing some fingers or adding some visual effect On the sign language to distort the language.

Discussion of the third question: What is the level of hearing impairment for Deaf people with Waardenburg syndrome type I on a Jordanian sample?

The results of the study showed that the degree of moderate hearing impairment was the highest category.

The researcher explains: Because of the auditory sensory loss of deaf people with Waardenburg syndrome type I and associated with the inner ear, which is the feature of this syndrome, all of its categories have a hearing loss of medium degree, and this indicates the use of sign language instead of lip

language due to the proportion of auditory loss for that category.

Recommendations:

Based on the findings: Educational recommendations:

1- Inclusion of this syndrome as an important part of the categories of hearing impairment.

2- Training for deaf workers on the characteristics of the Waardenburg type I syndrome.

3- Training workers to know the visual problems resulting from eye pigmentation, which may add shapes that are not present.

Research Recommendations:

1- Carrying out more studies and research related to Waardenburg syndrome of the first type for the hearing impaired.

2- - Conducting studies related to the comparison between deaf people with Waardenburg Syndrome and deaf people with intellectual disabilities.

3- Carrying out studies related to mental disorders in the Deaf with Type I syndrome.

4- Carry out studies related to visual perception in Deaf people with Type I syndrome.

5- Carrying out studies related to the level of intelligence of the deaf people with Type I syndrome.

Conclusion: The lack of researchers specialized in sign language at the level of translation of the deaf language makes it difficult to know and discover other cases accompanying hearing disability (such as double disability), and therefore there are many other cases that must be known in order to identify areas and levels within the category of hearing disability, which helps us to enrich this field Hence the secret in this scientific research lies in knowing the characteristics and rules of the sign language for the deaf in the researcher due to the ease of identification and access to other cases that do not appear with the normal measuring tools for that category.

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