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Impact of retinitis pigmentosa on daily and social life

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Abstract. Retinal dystrophies cause progressive degeneration of the retina in people who suffer from them, leading to loss of vision over the years that can negatively affect functional capacity and social participation.

Our study aims to analyse how ocularcentrism contributes to aggravating the negative consequences of retinal dystrophies for the daily and social lives of the population who suffer from these disorders in Castile and León.

This research used a mixed methodology with a QUAN+QUAL design. The quantitative part consisted of a case-control study with a non-probabilistic, purposive sample of 30 people with and without retinitis pigmentosa matched by age and sex. The World Health Organization's Model Disability Survey, Brief Version (MDSb) was administered to all participants. The study compared external factors that can influence daily health-related problems, family and social support, and attitudes of others. The qualitative part consisted of the analysis of data obtained through in-depth interviews with the 15 people with retinitis pigmentosa in the case group.

Significant differences were found between the groups, showing that people with dystrophies experience greater difficulties in their daily life than individuals who do not suffer from these disorders in areas such as mobility, household tasks and social participation. Noise, brightness, lack of technological means to facilitate accessibility and ocularcentrism in society represent a source of social exclusion, in addition to causing discomfort and tension.

The results provide evidence that will enable social workers to intervene in order to promote the social participation of a population affected by a rare eye disorder, thereby preventing inequalities and advancing their rights.

Keywords. Social participation; retinal dystrophies; daily life; quality of life.

[es] Impacto de la retinosis pigmentaria en la vida diaria y social

Resumen. Las distrofias retinianas, producen en las personas que las padecen, una progresiva degeneración de la retina que conlleva pérdida de la visión a lo largo de los años, afectando negativamente a su capacidad funcional y su participación social. Nuestro estudio pretende analizar cómo el ocularcentrismo, contribuye a agravar las consecuencias negativas que las distrofias retinianas producen en la vida diaria y social de las personas de Castilla y León que las padecen.

La investigación empleó metodología mixta con un diseño CUAN+CUAL. La parte cuantitativa fue un estudio de casos y controles con muestra no probabilística e intencional de 30 personas, con y sin retinosis pigmentaria pareados por edad y sexo. A todos ellos se les pasó la Encuesta Modelo de Discapacidad, Versión Corta (EMDc) de la Organización Mundial de la Salud. Se compararon factores externos, que pueden influir sobre problemas cotidianos relacionados con la salud, apoyo familiar y social y actitudes de los demás. La parte cualitativa consistió en el análisis de datos obtenidos mediante entrevistas en profundidad a las 15 personas con retinosis pigmentaria del grupo caso.

Se obtuvieron diferencias significativas entre los grupos, que muestran mayores dificultades de las personas con distrofias en su vida cotidiana que las que no la padecen, como la movilidad, la realización de tareas domésticas o con la participación social. El ruido, la luminosidad, no disponer de medios tecnológicos que faciliten la accesibilidad y el ocularcentrismo existente en la sociedad, además de provocarles malestar y nerviosismo, son fuente de exclusión social.

Los resultados aportan evidencias que permitirán a los/las trabajadores/as sociales intervenir para favorecer la participación social de un colectivo afectado por una enfermedad rara de la visión evitando inequidades y avanzar en sus derechos.

Palabras clave: Participación social; Distrofias retinianas; vida cotidiana; calidad de vida.

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Introduction

Visual impairment is a major health problem. In fact, the World Health Organisation (WHO) estimates that 1.3 billion people worldwide were affected by some form of visual impairment in 2018. Most of these eye conditions (around 80%) can be prevented or cured (WHO, 2013). The increase in the number of people affected across the globe has recently become known, and it is estimated that in 2020 the number of people affected was at least 2.2 billion (WHO, 2020). These data involve certain nuances depending on the degree of visual impairment. In this regard, Rao (2019) indicates that, in 2010, about 32.4 million people were considered to be blind, while about 191 million had moderate or severe visual impairment (MVI). Moreover, the largest proportion of the affected population was aged 50 and over. That is, 84.6% of blind people were in this age range, while 77.5% had MSVI. This author also shows that there is a strong geographical gap in the incidence of this type of eye condition. In fact, the prevalence is 25 times higher in areas of the globe with fewer resources. So much so that in North America the prevalence is 0.3%, while in South Asia it reaches 8.3%. Furthermore, 90% of people affected by pathologies linked to low vision or blindness are found in population groups with fewer resources (Rao, 2019). Over time, cases of blindness have also increased. Bourne et al. (2017) indicate that in 2015, it was estimated that 36 million people worldwide were blind, while 217 million had some form of moderate or severe visual impairment and, finally, 188 million people were affected by mild visual impairment.

Current data show that in 2018 there were 188.5 million people with moderate visual impairment, and 217 million with moderate to severe visual impairment. This information reveals that, in that year and across the world, there were approximately 405.5 million people affected by what we call Low Vision (LV). LV is, therefore, a condition caused by some form of pathology or causal accident related to vision (WHO, 2020). In short, low vision is a concept equivalent to moderate or severe visual impairment. The International Classification of Diseases version 11.a states that visual function can be categorised according to the level of difficulty in seeing. On the basis of this classification, we can rate individuals as having no visual impairment, a moderate visual impairment, a severe visual impairment and blindness. Low vision, therefore, encompasses the two intermediate categories.

Low vision is a human condition related to the reduction of people's well-being and, therefore, their quality of life; which brings about a reduction in the social participation of these individuals (Shah et al., 2019) and even increases the likelihood of suffering burnout (Wrzesińska et al., 2019). Hence, following Archer (2016), this population is potentially on the periphery of the social structure and its morphology. This fact can be aggravated in regions with a higher degree of depopulation (which is associated with a lack of social and health services). For this reason, we have focused our interest on the Spanish region of Castile and León, in which there are numerous provinces with high levels of depopulation and lack of services. In order to know exactly how many people are potentially affected by LV and by its negative impacts, we must go back to the survey carried out by Spain's National Statistics Institute (INE) in 2008, as these are the most reliable and official data on the reality of low vision in Spain. According to this survey, in Spain there were 979,200 people over the age of six with visual impairments and deficiencies at that time. Of these, 37.93% were male and 62.07% female. According to the same source, and for that year, in Castile and León there were 69,100 people over 6 years of age with visual disabilities and impairments, of whom 39.22% were men and 60.78% were women. According to the same source, 36,100 people in this region lived in municipalities with less than 10,000 inhabitants, which indicates that more than half of the people affected by visual impairment experience, foreseeably, social and health difficulties.

However, there is an interesting paradox resulting from the efforts of related social groups. The paradox is that people with blindness are socially the most widely recognised visually impaired population. By contrast, low vision is a more unknown disability. In fact, both students and professionals in the social and health care field are not familiar with the concept of low vision and its implications (Del Álamo & Del Álamo, 2019). On the other hand, and regarding the process of morphogenesis of modern society in its different variants (early modernity and late modernity), we can affirm that today's society has been shaped by placing vision at the centre of its structure. For this reason, Bulk et al. (2020) refer to the concept of ocularcentrism to show that society establishes an all-or-nothing logic: vision/no vision. Hence, intermediate disabilities like low vision seem to be excluded from the social structure, making it difficult for them to participate in the generation and development of society.

In this paper we will focus our interest on people affected by retinitis pigmentosa, also known as pigmentary retinitis, and how ocularcentrism negatively affects their daily lives. This pathology is a rare disease whose prevalence, according to the Orphanet platform, is 1-5/10,000 people. When we talk about retinitis pigmentosa (RP) we refer to a group of genetic (hereditary) disorders that bring about a gradual progressive retinal degeneration. More specifically, it affects the photoreceptors and their pigment epithelium (García et al., 2020). RP is genetically very heterogeneous as mutations in more than 50 genes are known to cause non-syndromic RP and about 3100 mutations in these genes have been reported (Daiger et al., 2013). The clinical features of this condition are mainly night blindness, which is usually followed by progressive loss of peripheral vision, as well as loss of central vision (Daiger et al., 2013; Garcia, et al., 2020).

Our hypotheses are precisely related to this.

- *H1: Ocularcentrism excludes people with retinitis pigmentosa from the social structure.*
- *H2: Ocularcentrism implies a reduced participation of people with retinitis pigmentosa in the evolution of the social system.*

The present research has, therefore, the intention to analyse ocularcentrism as an element shaping social significance and to learn about the difficulties and situations that people with RP living in the region of Castile and León (Spain) have to face.

1. Metodology

The present study is of a mixed nature (quantitative and qualitative). However, it is descriptive and comprehensive in character, as well as presenting a case-control analysis. It was carried out from February to June 2020 with a case group made up of members of the Retina Castilla y León association affected by RP and a control group made up of people without RP. The quantitative research was completed with in-depth interviews conducted in person and by telephone with the participants of the case group. Sampling was non-probabilistic and intentional. Thirty people were recruited through telephone calls, 15 of whom were affected by RP and 15 of whom were not affected by RP. Calls were made until the groups were matched by age, sex and rural/urban origin. All the people resided in municipalities in the region of Castilla y León. All were informed of the purpose of the study as well as of its voluntary nature, and were guaranteed the confidentiality of the data and their anonymisation by means of informed consent.

1.1. Measuring instruments

The World Health Organization’s Model Disability Survey, Short Version (MDSc) (2018) was used for the purpose of this study. This survey (MDSc) is a shorter version of the Model Disability Survey (MDS), developed by WHO and the World Bank, in collaboration with other stakeholders in the disability field. It is based on the International Classification of Functioning, Disability and Health (ICF) and understands disability as “the result of an interaction between a person’s health condition(s) and the physical, human, attitudinal and socio-political environment in which he or she lives” (WHO, 2018).

The survey was designed with a threefold objective: “to provide detailed and nuanced information about the lives of people with disabilities; to enable comparison between groups with different levels and profiles of disability; and to provide the evidence needed by policymakers to identify what interventions are needed to optimise the inclusion and functioning of people with disabilities” (WHO, 2018).

The survey consists of several modules. The first of these relates to “Environmental Factors” broadly understood as all external factors that affect the person’s lived experience of disability. This module includes questions related to:

- Aspects of the environment that hinder or facilitate. These aspects would allow the identification of barriers or facilitators that may influence everyday health-related problems. The response options are on a scale of 1 to 5, where 1 means “very easy” and 5 means “very difficult”.
- Family and social support. As in the previous section, the response options are on a scale of 1 to 5, where 1 means “very easy” and 5 means “very difficult”.
- Attitudes of others. The questions are based on the WHOQOL-BREF Disability Module. Response options are on a scale of 1 to 5, where 1 means “no, not at all” and 5 means “yes, completely.

The second one refers to “Functioning” through 12 questions that ask how much of a problem it is to perform activities related to mobility, self-care, stress management, cognition, housework, community participation and employment or school. Therefore, “problems” are understood as the obstacles the person faces in performing activities. Obstacles may arise from their health, but also from the environment in which they live. The response options are also a Likert-type scale from 1 to 5, where 1 means not problematic at all and 5 means that the action is extremely problematic for them or they cannot do it. The survey also includes two more modules, referring to capacity and health conditions and to technical aids and personal assistant.

1.2. Data analysis

Quantitative data were analysed using SPSS software. First, for the quantitative variable age, a descriptive analysis of the sample was performed. Subsequently, in order to test the differences between the case group and the control group, the non-parametric Mann-Whitney U test was used for independent samples, as normal distribution was not confirmed by the Shapiro-Wilk test. Contingency tables and the chi-square test were used for qualitative variables. Subsequently, we proceeded to study the internal consistency of the instrument for the sample by means of Cronwach's alpha reliability analysis.

The data obtained from the interviews were analysed by conducting a content analysis. After the first general reading of the transcripts, a first identification of codes and categories was made with the participation of two members of the research team. In a first phase of analysis, a free coding of part of the transcribed material was carried out and in a second phase, with the participation of another member of the team, this initial coding was revised but now including key terms identified in the literature. This first confrontation of categories served to discuss and agree on the criteria for the semantic fields of interest and the coding guide itself. After the coding process, the results were structured into four semantic blocks: Social Niche, Technological Development, Exclusion/Unknown and Emotions/Nervousness.

2. Results

2.1. Socio-demographic characteristics of the case and control groups.

The sample (see Table 1) consists of 30 young adults of working age (86.7%) and older adults (13.3%). All the people in the sample are above the second decade of life. The mean age of the participants was 53.53 years in the case group, and the standard deviation of the case group was 10.54 years. By contrast, the control group has a mean age of 53.46 years and a standard deviation of 11.02 years. In both groups (case and control), the following variables were controlled: sex, age and residence. Eighty percent were male and 20% female, firstly because RP is more common in men and, secondly, because the association that collaborated in this study has a greater number of male than of female members. Both circumstances made it difficult for women to participate. On the other hand, an attempt was made to adjust the participation of people from rural and urban areas with the official data mentioned above. Hence, 60% of the participants live in rural areas and 40% in urban areas.

Tabla 1 .*Socio-demographic aspects of the participating groups*

Variables		With Hereditary Retinal Dystrophies (n=15)		No Hereditary Retinal Dystrophies (n=15)		Total (n=30)	
		F	%	F	%	F	%
Age	20-40	1	6.7	1	6.7	2	6.7
	41-60	10	66.7	10	66.7	20	66.7
	61-80	4	26.7	4	26.7	8	26.7
Sex	Woman	3	20	3	20	6	20
	Man	12	80	12	80	24	80
Origin	Rural	6	40	6	40	12	40
	Urban	9	60	9	60	18	60

Source: own elaboration

In order to fulfil the objective of finding out if there were significant differences in age depending on whether they had RP or not, after performing the Shapiro-Wilk normality test on the case (p=.963) and control (p=.704) groups, the Mann-Whitney U test was carried out and it was found that there were no differences between the groups. Table 2 shows the results.

Tabla 2. *Differences between having retinitis pigmentosa or not according to age.*

	With Hereditary Retinal Dystrophies (n=15)	No Hereditary Retinal Dystrophies (n=15)			
	Average range	Average range	Z	U	p

Age	15.40	15.60	-0.62	111	.967

Source: own elaboration. Note:*$p<0.05$

Table 3 shows the characteristics of the interviewees.

Tabla 3. *Characteristics of the participants interviewed*

Participating subjects	Age	Origin	Illness
Man 1	32	Urban	RP
Man 2	51	Urban	RP
Man 3	46	Rural	RP
Man 4	42	Urban	RP
Man 5	59	Rural	RP
Man 6	72	Rural	RP
Man 7	63	Rural	RP + Usher syndrome II
Man 8	57	Urban	RP
Man 9	66	Rural	RP
Man 10	48	Rural	RP + Hearing loss
Man 11	60	Urban	RP
Man 12	49	Urban	RP
Woman 1	50	Urban	RP
Woman 2	63	Urban	RP
Woman 3	45	Urban	RP + Magna Myopia

Source: own elaboration

2.2. Results of the MDSc

The validity and reliability of the short version of the MDSc were acceptable, with a Cronbach's alpha value of 0.746. The subscale "Environmental factors" was also acceptable. (see Table 4).

Tabla 4. Valor de Alfa de Cronbach si el ítem es excluido

Ítems	**Alfa deCronbach según pregunta excluida**
¿En qué medida los lugares donde usted socializa y participa en actividades de la comunidad le hacen fácil o difícil hacerlo	,764
¿En qué medida las tiendas, los bancos y la oficina de correo de su vecindario le hacen fácil o difícil utilizarlos?	,547
¿En qué medida los medios de transporte que usted necesita o quiere usar le hacen fácil o difícil utilizarlos?	,547
Si necesita ayuda, ¿en qué medida le resulta fácil o difícil conseguir ayuda de un pariente cercano (incluido su cónyuge)?	,549
Si necesita ayuda, ¿en qué medida le resulta fácil o difícil conseguir ayuda de amigos o compañeros de trabajo?	,763
Si necesita ayuda, ¿en qué medida le resulta fácil o difícil conseguir ayuda de vecinos?	,762
¿Puede tomar sus propias decisiones en relación con su vida diaria? Por ejemplo, a dónde ir, qué hacer, qué comer.	,767
¿Siente que las demás personas lo respetan? Por ejemplo, ¿siente que lo valoran como persona y que escuchan lo que tiene que decir?	,767
Total	**,746**

Tables 5 and 6 present the comparison of the groups with and without hereditary retinal dystrophies on the dimensions environmental factors and functioning.

Tabla 5. *Tabla de contingencias de la dimensión factores ambientales y comparación entre tener distrofias hereditarias de retina y no tenerlas*

	Con Distrofias Hereditarias de Retina					*Sin Distrofias Hereditarias de Retina*								
Módulo 3 Factores externos	*Muy fácil*	*Fácil*	*Normal*	*Difícil*	*Muy Difícil*	*Muy fácil*	*Fácil*	*Normal*	*Difícil*	*Muy Difícil*	TOTAL	X2	gl	p
¿En qué medida los lugares donde usted socializa y participa en actividades de la comunidad le hacen fácil o difícil hacerlo?	1	2	6	6	0	14	1	0	1	1	30	23,600^a	3	*,001**
¿En qué medida las tiendas, los bancos y la oficina de correo de su vecindario le hacen fácil o difícil utilizarlos?	0	6	4	5	0	12	1	0	1	1	30	23,238^a	4	*,001**
¿En qué medida los medios de transporte que usted necesita o quiere usar le hacen fácil o difícil utilizarlos?	1	2	6	5	1	9	2	2	0	1	30	14,400^a	5	*,013**
Si necesita ayuda, ¿en qué medida le resulta fácil o difícil conseguir ayuda de un pariente cercano (incluido su cónyuge)?	11	3	0	1	0	8	3	3	0	0	30	5,474^a	4	*,242*
Si necesita ayuda, ¿en qué medida le resulta fácil o difícil conseguir ayuda de amigos o compañeros de trabajo?	11	4	0	0	0	0	0	0	1	14	30	30,000^a	3	*,001**
Si necesita ayuda, ¿en qué medida le resulta fácil o difícil conseguir ayuda de vecinos?	8	1	3	2	0	0	0	0	4	11	30	24,667^a	5	*,001**
¿Puede tomar sus propias decisiones en relación con su vida diaria? Por ejemplo, a dónde ir, qué hacer, qué comer.	0	0	0	0	15	15	0	0	0	0	30	30,000^a	1	*,001**
¿Siente que las demás personas lo respetan? Por ejemplo, ¿siente que lo valoran como persona y que escuchan lo que tiene que decir?	0	0	0	1	14	14	1	0	0	0	30	30,000^a	3	*,001**

Fuente: elaboración propia. Nota:*p<0.05

Tabla 6. *Tabla de contingencias del funcionamiento y comparación entre tener distrofias hereditarias de retina y no tenerlas*

	Con Distrofias Hereditarias de Retina					*Sin Distrofias Hereditarias de Retina*								
Módulo 4 Funcionamiento	*Nada*	*Poco*	*Bastan-te*	*Mucho*	*Extremo*	*Nada*	*Poco*	*Bastan-te*	*Mucho*	*Extremo*	TOTAL	X2	gl	p
¿Qué tan problemático es para usted llegar a los lugares a donde quiere ir?	0	6	4	5	0	12	1	0	1	1	30	26,250	3	*,001**
¿Qué tan problemático es para usted asearse y vestirse?	12	1	2			15					30	3,333a	2	,189
¿Qué tan problemático es para usted usar el baño?	1	2	6	5	1	9	2	2	0	1	30	5,138a	2	,077
¿Qué tan problemático es para usted cuidar de su propia salud, comer de manera saludable, hacer ejercicio o tomar sus medicamentos?	11	3	0	1	0	8	3	3	0	0	30	6,303a	2	,043
¿Qué tan problemático es para usted sentirse cansado y no tener suficiente energía?	11	4	0	0	0	0	0	0	1	14	30	6,000a	3	,112
¿Qué tan problemático es para usted hacer frente a todas las cosas que usted tiene que hacer?	8	1	3	2	0	0	0	0	4	11	30	2,571a	2	,276
¿Qué tan problemático es para usted recordar las cosas importantes que tiene que hacer en su vida diaria?	0	0	0	0	15	15	0	0	0	0	30	11,763a	2	*,003**
¿Qué tan problemático es para usted hacer las tareas de la casa como barrer, cocinar o sacar la basura?	5	4	5	0	0	14	1	0	0	0	30	12,063^a	3	*,007**
¿Qué tan problemático es para usted participar en actividades de la comunidad, como fiestas, eventos religiosos u otras actividades	3	3	6	3	0	14	1	0	0	0	30	17,118^a	3	*,001**
¿Qué tan problemático es para usted utilizar servicios de transporte públicos o privados?	2	5	6	2	15	0	0	0	0	0	30	22,941^a	3	*,000**

Fuente: elaboración propia. Nota:*p<0.05

In response to the question "To what extent do the places where you socialise and participate in community activities make it easy or difficult for you to do so?" 40% of the case group considered that these places make it difficult to socialise and participate, compared to 86.67% of the control group who reported the opposite situation. The chi-square test (X2=23.60; gl=3; p=0.000*) allows us to affirm that the perception of greater difficulty in carrying out leisure activities is associated with having a visual impairment. Something similar occurs with regard to access to and use of bureaucratic and institutional places. For 86.67% of the participants in the control group it was a very easy task, in contrast to 33.33% of the case population, who found it very difficult. In this case, the chi-square test (X2=23.238; gl=4; p=0.000*) allows us to affirm that the perception of greater difficulty in using places such as shops, banks and post offices is associated with having a visual impairment. In the case of access to public or private transport, 93.33% of the control group found it very easy, while 33.33% of the case group found it difficult and 6.67% very difficult. The chi-square test (X2=14.40; gl=5; p=0.013*) allows us to affirm that the perception of greater difficulty in using public or private means of transport is associated with having a visual impairment.

When asked about how problematic it was for respondents to get to where they wanted to go, 46.67% of the case group stated that this process was very problematic. Likewise, 20% of this same group responded that this problematicity was extreme. On the other hand, 93.33% of the control population found this action very easy. The chi-square test (X2=26.25; gl=3; p=0.001*) allows us to state that there is a clear association between the difficulty of human movement and the perception of greater difficulty in getting to where one wants to go or where one has to go.

In relation to the degree of difficulty perceived by the participants while performing household tasks with some kind of support, 93.33% of the control group claimed that this situation does not pose any difficulty. On the other hand, in the case population, there is a greater perception of difficulty, as 33.33% associate these tasks with quite a lot of difficulty. The chi-square test (X2=12.06; gl=3; p=0.007*) that was carried out allows us to establish a clear association between visual impairment and the perception of greater difficulty in performing household tasks (even with support). When we ask about the performance of the same tasks without any help, the data, in percentage terms, are similar. However, the chi-square test (X2=19.67; gl=3; p=0.000*) increases.

Another aspect of great relevance is participation in social activities (parties, events, etc.). When asked about the degree of difficulty experienced in taking part in this type of activities (with help), we found that 86.67% of the control population perceived them as an easy activities, while 40% of the case group found them,quite difficult and 20% very difficult. Therefore, the chi-square test (X2=13.82; gl=3; p=0.003*) shows that there is an association between the perception of greater difficulty in participating in social activities and visual impairment. Finally, when asked about the perceived difficulty of this same type of social activities (without support), we see that 26.67% of the case group perceives it as quite difficult. By contrast, 93.33% of the control population responded that, for them, such activities were not difficult at all. The chi-square test (X2=20.00; gl=3; p=0.000*) allows us to affirm that the perception of greater difficulty in participating in community activities in an autonomous manner is associated with having a visual impairment.

2.3. Qualitative Results.

The qualitative analysis enables us to structure the responses of informants into the following semantic fields: Social niche, Technological development, Exclusion/unawareness and Emotions/nervousness. The first semantic field is possibly the most obvious, as it refers to difficulties related to physical accessibility: stairs, poorly signaled glass panels, noise, etc. In this sense, 73% of informants indicate that noise hinders their perceptive capacity, as they do not have the visual option to manage their activity in the social structure. On the other hand, brightness is also important for those affected, since not all people affected by RP (as we indicated at the beginning) are blind. In our research 93% were not blind.

The second semantic field referred to by informants is that of technological development. The non-existence of technology-mediated accessibility mechanisms hampers their capacities. The social structure presupposes the ability to see and eliminates alternatives to this structural preconception. In fact, one of the participants stated: "*I went to the administration, there I had to fill in a form, but my RP is already advanced and it was impossible because the letters were too small*" (Man 6).

Once again we find this ocular centrality reducing the options for people with low vision. Related to this, in the interviews we were told of difficulties in locating bus card readers (40% of informants). In this sense, the informants emphasise the importance of being acquainted with the environment and, therefore, the need for the social niche to be structured on the basis of the potential lack of knowledge on the part of the population.

The third semantic field, that of exclusion/unawareness, was relevant in the interviews. One of the collaborators stated: "*I asked a lady at the counter and she answered: 'Are you blind? Can't you do it on your own? It was quite humiliating*" (Woman 1). Another indicated: "*I was going to get on the bus, but there were people behind me and I got very nervous, I asked the bus driver if he could show me where the card machine was and he kept saying 'there', which only made me more nervous; finally he said 'can't you see it?', which was indeed true*" (Woman 3). This shows the effects of social ignorance and the phenomenon of what could be called a zero-level exclusion, in which there is no intention to exclude, but the social structure and the normalising conception of reality (if we may use the term) generate such exclusion. It is here again where not only the barriers of the environment, but also the ignorance of society, hinder the social participation of people with LV.

The fourth semantic field is that of emotions. The lack of knowledge of the environment and the difficulties associated with low vision generate emotional pressure and nervousness in these persons. One of the informants, in this sense, claims: "*my wife/daughter/brother is my hands, she has to be constantly supervising what I do, so that I don't keep dropping things*" (Man 7). In addition, the mere fact of accessing any place, whether it is known or unfamiliar, can be a challenge for people with low vision. Therefore, it is essential —as shown in the first semantic field— to establish a social niche that is as accessible as possible and where ocularcentrism is not so important. In fact, when asked about participation in social activities, informants with RP (93%) pointed out that the difficulty of finding a person to accompany them limits their socialisation.

3. Discussion

Everyday life is about social interactions and human agency, that is to say, with the capacity that people have to develop activities of a collective nature. However, talking about agency implies talking about ontological aspects of "humanity" and "sociality". Margaret S. Archer (2009) in her book *Being Human* studies this aspect from an intermediate perspective between the idea of "Modern Man" and "Social Being". The former, she argues, is based on the assertion that society does not contribute to our own configuration. The latter, on the other hand, is established on the basis of the opposite view: society contributes completely and totally to our own personal configuration. Archer (2009) argues, then, for a stratified view of the "subject". As the subject moves between different strata, different properties and powers will emerge at each level. She differentiates between four strata: the self, the person, the agent and the actor. Each level possesses differential characteristics. Thus, the level of the self will be confined to the private sphere and will have a type of individual realisation. The personal level corresponds to a type of primary agency, with a private character and a social or collective realisation. The agent level implies a level of collective realisation, an eminently public character where the person enters groups. These social groups will contribute to the process of reproduction and change of the social structure. In this sense, Archer (2009) goes so far as to consider these agents as corporative. Lastly, we come to the actor. This has a triple character, since it is made up of individuals and social groups (and therefore has also a public character). But, in addition, these individuals and social groups are structured in organisations and, therefore, fulfil an organisational role that the previous strata do not have. In this way, Archer (2009) structures a continuum through which an interrelationship is established between different levels of agency and human activity, whether individual or collective.

This continuum has an important heuristic potential to explain the processes we have been studying and to understand the mechanisms of (dis)structuring to which people with retinitis pigmentosa (RP) are subjected. From this structural perspective, the influence that the public dimension has on the private dimension and vice versa is clear. If a person with RP (or low vision in general) cannot become a social actor and is forced to reduce his or her participation, his or her own self is affected. This was recently verified by Fraser et al. (2019) who stated that the effects on all these levels create barriers to socialisation in older people with low vision. This has negative effects on the well-being, health and quality of life of persons with low vision (Shah et al., 2020; Garip and Kamal, 2019). In this sense, if we take into account that in the interviews conducted, all the participants stated that RP had negatively influenced their social participation, we can argue that this biomedical element (RP) reduces their capacity for social agency, together with the perception and relationships that others have with people affected by RP. However, people with RP have to struggle to maintain and increase their level of social agency. Currently, the process of institutionalisation has made it easier for them to build their capacity for agency, reaching the level of social actors. However, we have seen that the demand for the development of the individual self is still far from adequate.

From the moment RP manifests itself, people are conditioned by the processes of adapting their self to the new reality generated by the pathology. In addition, they also have to deal with stigmatisation, differential treatment and denial of opportunities (Bulk et al., 2020; Sivakumar et al., 2020). These processes can be produced by the affected subjects themselves or by others. However, Petereit et al. (2019) have found that social exclusion produces biological effects, generating changes in oxytocin production, which affects the neural processing of the feeling of rejection. This leads us to think that biosocially, social exclusion (heterogenerated) is more important than self-generated exclusion. However, much remains to be known about the interaction of oxytocin with other neurotransmitters (e.g., acetylcholine, GABA, dopamine, etc.) to modulate social neural signals (Maltese and Papaleo, 2020).

The phenomenon of exclusion is often mediated by stigmatisation processes, which devalue some factor of some group in certain social contexts (Fraser et al., 2019). However, there is a process of unconscious exclusion that can be referred to as zero social exclusion in which the person generating exclusion does not realise that he or she is carrying out such a process. This fact was evinced by our informants when they mentioned, in the interviews, that people take for granted that we all see equally well. However, there is also a process of overt social exclusion. In fact, visually impaired people show a significantly lower participation in social activities and have high rates of social isolation (Bulk et al., 2020). For example, many of the participants, although already retired due to their degree of disability, reported problems, not only socially but also at work, in the early stages of diagnosis of the disease.

A large part of the social barriers affecting people with RP (and, of course, people with low vision in general) lie in the physical structure of the social niche. In this sense, our work complements that of Senthil et al. (2017) where the process of exclusion of these individuals was not analysed. A phenomenon that, on the other hand, is in line with the data studied in relation to people with low vision (Fraser et al., 2019). The element that allows us to understand the destructuring mechanism to which persons with RP are subjected is that of ocularcentrism: a concept that Moya et al. (2020) have studied in order to show how the city has been structured, since Modernity, on the basis of visual pre-eminence, relegating people with different sensorialities to the margins of the social structure. We are therefore considering the need to establish permanent and compulsory training mechanisms in the world of work. This may require a legal framework similar to the Law on the Prevention of Occupational Risks, i.e. a law that would promote citizenship awareness of the social reality we are discussing and would mitigate the negative effects on the individual (self) and personal development of this population (and of any other population affected by a disability).

Without explicitly mentioning it, Fraser et. al. (2019) showed this phenomenon by pointing out that shops, banks, hospitals, etc. are architecturally structured on the basis of this ocularcentrism. This creates artificial barriers for people with RP. In this sense, and as part of our quantitative results, the informants reported the greatest difficulty in accessing different places, while in the interviews they showed that they had often found themselves in complicated situations. One of the main mechanisms for reducing this process of destructuring undergone by this population is related to the acquaintance and familiarisation with the social niche (with the environment). This enables the other sensory capacities to become operational and helps in the processes of social participation and agency (Low, 2015). However, we should recall that Le et al. (2020) show a clear correlation between PR and anxiety and depression. This seems to call into question the benefits of familiarity as a mechanism for effectively reducing the negative effects of illness and ocularcentrism. In this sense, once again, we wonder whether a greater understanding of the social reality of this population group could engage architects and engineers in developing alternatives in the design of cities, buildings, etc.

Throughout this paper, we have shown different quantitative and qualitative data that allow us to infer the visual structuring of society. This forces persons with low vision, and more specifically those with retinitis pigmentosa, to establish themselves on the limits of the social structure, i.e., at the sites in the structure where there are limitations of social agency. Bulk et al. (2020) also found this to be true to some extent in blind people by showing the reduction in life expectancy of this population group. The reduction of social agency can be seen in the existing geographical barriers, as we showed in the results section , but there are also other elements that condition the development of agency. One of these conditional factors is intersubjective relationships. In fact, as the informants have shown, the immediate environment of persons affected by RP has a negative effect on the acceptance of the pathology or even on the adaptation to the new reality. This can lead to the social isolation and disintegration of the affected person, which requires greater support from the health, educational and social systems (Bertelsen et al., 2015).

On the other hand, it is very enlightening to understand that a second element of exclusion to which people with RP are subjected is linked to the interpretative prejudice of blindness. In other words, the current social structure interprets vision problems on the basis of the classic logic: all or nothing. Thus, Fraser et al. (2019) indicate that the population does not differentiate between partial or absolute vision loss. In fact, some perspns with low vision do not need aids such as a cane or a guide dog. This causes them to be socially interpreted as people without vision problems, which can be problematic. In addition, the negative emotions and preconceptions of people with RP (fear of rejection, doubts about other people's reactions, mistrust, etc.) together with the need to feel "normal" often make people with RP want to go unnoticed. This fact has also been observed in other types of rare diseases (Coca, 2019) and it can generate additional problems such as the one mentioned above regarding the detection of the card reader on the bus.

As limitations to our study, we could point out a bias in the selection of the case group because they are all members of the same association. This bias could have been avoided if the sample had also been taken from ophthalmology practices in different health areas, but the limitations imposed on fieldwork during the covid-19 pandemic prevented us from doing so. On the other hand, the scarcity of existing literature on the impact of RP on the daily and social life of people who suffer from this condition in the field of social sciences, and in particular in social work, has prevented us from comparing our results with other studies of similar characteristics.

4. Conclusions

Retinitis pigmentosa is a rare disease that generates what is known as low vision. This condition entails a social and personal reality that is different from that of blind people and, of course, also from that of people without serious vision problems. In this paper we have seen how one of the major factors that exclude people with low vision from the structure of society is the centrality of vision in the configuration of the social system itself. All social reality starts from the premise, whether consciously or not, that people are capable of visually perceiving reality. This fact forces persons with LV into a marginal position, limits their lives and even substantially reduces their capacity for social agency. Throughout this paper we have shown how people with retinitis pigmentosa in the region of Castile and León suffer from a double marginalisation due to the absence of differentiating elements of their visual impairment. This can be explained, once again, by the ocularcentrism that operates in society. In view of this fact, it is essential for the affected individuals and groups (social actors) to demand that state institutions increase their capacity for social agency so that they are ultimately able to live socially and with the greatest possible degree of equity. In this sense, we argue for the need to establish job training mechanisms to reduce the existing exclusion gap. The description of the social reality shown in this research evidences the role that social work should play as a mediator for the social inclusion of people with RP (Díaz-Jiménez et al., 2021; Munuera, 2015) and as a facilitator of social participation by generating, through community action, collective dynamics that allow for the construction of a civic culture of the collective space from a social rights approach (Camps, 2000; Moya et al., 2020). Our findings open the gateway to future social research so as to reduce or transform the ocular paradigm and highlight the need for social policy makers to take further steps in order to develop relevant and effective interventions leading to the removal of the stigmatisation of visually impaired citizens. .

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