

# Causes and characteristics of low vision patients in Turkey

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## 土耳其低视力患者的病因及特征

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### 摘要

**目的:**探讨土耳其配戴助视器的低视力患者的病因和特点,及其与父母近亲结婚是否有关系。

**方法:**回顾性研究了2009/2013年间伊斯坦布尔诊所接受236例低视力患者。分析年龄、性别分布、低视力原因、近亲结婚、低视力辅助类型规定及视力变化。在回顾性研究中,根据世界卫生组织定义基于最佳矫正视力将视力分为(失明,视力 $<20/400$ ;视力严重受损,视力 $<20/200 \sim 20/400$ ;视力轻度到中度受损,视力 $<20/60 \sim 20/200$ )。基于双眼主要病因,根据国际疾病分类第十版可以确定失明和低视力的病因。根据视力和患者需求规定了视力辅助,并使用平均值 $\pm$ 标准差和频率对数据进行了描述。

**结果:**研究包括236例患者,65%为男性,平均年龄为 $38.5 \pm 24.2$ 岁(年龄范围6~95岁),男性多于女性。年龄在15~30岁(35.6%)之间是最大的年龄群体。轻度至中度视力受损患者为122例,严重视力受损患者为84例,失明患者为30例,他们分别占总人数的51.6%、35.6%和12.7%。脉络膜疾病和视网膜疾病(62.7%)是导致视力低下的主要原因。老年低视力患者黄斑变性逐渐成为视力低下的首要原因(61.3%)。造成视力低下的原因有视网膜和脉络膜疾病(62.7%),眼球震颤(23.7%),视神经和视神经束疾病(11%),先天性白内障(0.8%)及青光眼(1.7%)。88例患者(37.3%)为伽利略类型,116例患者(49.2%)为开普勒类型。18例患者(7.6%)在接受LVA后视力无明显改善。14例患者被准许使用放大镜矫正视力。在大多数患者中,使用LVA的近视和远视患者视力均得到改善。62例近亲结婚患者占总人数的26.3%。

**结论:**14岁以上患者视力受损主要原因是视网膜和脉络膜疾病,15岁以下的患者常见原因为眼球震颤。在近亲结婚群体中黄斑营养不良和色素性视网膜炎患者显著增高。研究疾病病因学的遗传非常有效,因此,近亲结婚在土耳其仍是一大问题。

**关键词:** 视力低下;近亲结婚;低视力辅助

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### Abstract

• **AIM:** To describe causes, characteristics and parental consanguineous marriage of low vision patients referred to our clinic for taking low vision aids in Turkey.

• **METHODS:** We conducted a retrospective study of 236 low vision patients who visited to our clinics in Istanbul from 2009 to 2013. Age and sex distribution, cause of low vision, parental consanguineous marriage, type of prescribed low vision aids, and changes of the visions were reviewed. In this retrospective study, visual acuity was classified based on best-corrected visual acuity in the better eye according to the World Health Organization definition (blindness, visual acuity (VA) $<20/400$ ; severe visual impairment, VA $<20/200 - 20/400$ ; mild to moderate visual impairment, VA $<20/60 - 20/200$ ). The causes of blindness and low vision were determined using the 10th version of International Classification of Diseases based on the main cause in both eyes. Vision aids were prescribed based on visual acuity and patients' requirements. To describe data, we used mean $\pm$ SD and frequency.

• **RESULTS:** The study included 236 patients, 65% male, with a mean age of  $38.5 \pm 24.2$  years (range, 6 to 95 year). In result, male were more than female. The age group between 15 and 30-year-old (35.6%) was the largest age group. Mild to moderate visual impairment, severe visual impairment and blindness were present in 122 (51.6%), 84 (35.6%) and 30 (12.7%) of the patients, respectively. Choroid and retina diseases (62.7%) were main causes of low vision. Elderly low vision patients macular degeneration is becoming a leading cause of low vision (61.3%). The causes of visual impairment were retinal and choroidal diseases (62.7%), nystagmus (23.7%), optic nerve and optic tract diseases (11%), congenital cataract (0.8%), and glaucoma (1.7%). Eighty-eight (37.3%) patients Galilean type; 116 (49.2%) patients keplerian type telescopic glasses recommended. Eighteen (7.6%) patients had no improvement in visual acuity with low vision aids (LVA). Forteen patients (5.9%) were approved the magnifier for near vision. In most patients, the use of LVA is improved both near and distance visual function. Parental consanguinity is present in 62 (26.3%) patients, most commonly between the ages of 15-30 were significantly higher in the group (50%).

• **CONCLUSION:** Diseases of the retina and choroid are the main cause over 14 years old groups, nistagmus is the most common cause under 15 years old group of visual impairment among patients referred to our clinic in Turkey. Parental consanguinity was significantly higher in macular dystrophy and retinitis pigmentosa groups. Of genetics in the etiology of these diseases are known to be effective. For this reason, parental consanguineous marriage is a big problem still in Turkey.

• **KEYWORDS:** low vision; parental consanguineous marriage; low vision aid

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## INTRODUCTION

According to a World Health Organization (WHO) consultation report, person with low vision is someone who has; an impairment of visual function, even after treatment or refractive correction; a visual acuity of less than 6/18 (20/60) to perception of light or a visual field of less than 10° in the better eye, but who uses, or is potentially able to use, vision for the planning and/or execution of a task<sup>[1]</sup>. Low vision is one of the priorities in the global initiative, VISION 2020 – The Right to Sight, along with cataract, trachoma, onchocerciasis, childhood blindness, and refractive error<sup>[2]</sup>. Low vision and blindness are two important health and socioeconomic issues in developed and developing countries. Demographic conditions, socioeconomic status and cultural differences have a deep impact on the prevalence and distribution pattern of this disability. Additionally, the causes of their incidence in different ages vary and some of them are preventable<sup>[3-5]</sup>. Therefore, epidemiologic evaluation and examining the causes of visual impairment in any society is a matter of concern and has a direct effect on the country's health care planning<sup>[6-8]</sup>.

Patients with visual impairment require special eye care including clinical evaluation, consultation and rehabilitation to improve quality of life and decrease their dependency<sup>[9-13]</sup>. In previous studies, the prevalence of low vision in patients older than 75 years was 10.3% and increased rapidly by age to about 30% at the ages of 90 years and above<sup>[14]</sup>.

In this study, therefore, we investigated characteristics of low vision patients such as age and sex distribution, cause of low vision, parental consanguineous marriage, type of prescribed low vision aid in Turkey. We expect these data can be useful for planning low vision services, active care and rehabilitation.

## SUBJECTS AND METHODS

**Subjects** This descriptive study was conducted on completely blind and visually-impaired patients who presented our clinic to receive visual aids. All patients provided written informed consent. Patients were examined by same ophthalmologist. At the end of the examination, vision aids were prescribed based on visual acuity and patients' requirements. The

**Table 1** Age, sex, parental consanguineous marriage distribution of low vision patients

Age (a)	Patientno.	Sex (M)	Consanguineous marriage n(%)
<15	34 (14.4)	20 (58.8)	6 (17.6)
15-30	84 (35.6)	60 (71.4)	42 (50) <sup>b</sup>
31-50	50 (21.2)	30 (60)	10 (20)
>50	68 (28.8)	46 (67.6)	4 (5.9) <sup>b</sup>
Total	236	156 (66.1)	62 (26.3)

<sup>b</sup>P<0.001.

vision aids consisted of typical far glasses, telescopic glasses (power 2× and 4×), max TV (power 2×), hand-held magnifiers (with or without illumination).

**Methods** Refractive error was measured using an autorefractometer. Best-corrected visual acuity (BCVA) and uncorrected visual acuity (UCVA) were assessed using the Snellen chart. Near vision was assessed using the Amsler chart. Patients were divided into three groups according to their BCVA following WHO classification method: 1) mild to moderate low vision (20/60 < BCVA ≤ 20/200), 2) severe low vision (20/200 < BCVA ≤ 20/400), and 3) blind (BCVA < 20/400)<sup>[7]</sup>.

All physicians who referred the patients were asked to describe the main cause of low vision based on their clinical and paraclinical evaluations. For any referred patient without an exact diagnosis, further evaluation and diagnostic procedures were recommended and patients were referred to an expert ophthalmologist.

The International Classification of Diseases (ICD-10) was used to classify the patients according to the primary cause of low vision and blindness for both eyes<sup>[8]</sup>. Based on this classification, any disease is categorized by a specific code which itself is a subheading of a general heading that classifies the disorders based on the anatomic site of involvement.

**Statistical Analysis** To describe data, we used mean ± SD and frequency (percent). All statistical analysis were performed using number cruncher statistical system (NCSS) 2007 & power analysis and sample size (PASS) 2008 statistical software (Utah, USA) programme.

Qualitative evaluation of data, Pearson's chi-square test to compare the data, Yates continuity correction test and Fisher's exact test was used.

## RESULTS

During the study period, 236 consecutively referred patients including 156 men (66.1%) and 80 women (33.9%) were enrolled. All were Turkish. The age ranged between 6 to 95 years old and the mean age was 38.5 ± 24.2 years. The largest age group was those between 15 and 30 years of age (35.6%), followed by over 50 years of age (28.8%), and 31-50 years of age (21.2%). Parental consanguinity was present in 62 (26.3%) patients, most commonly between the ages of 15-30 were significantly higher in the group (P < 0.001). Parental consanguinity over the age of 50 was significantly low (P < 0.001) (Table 1).

**Table 2 Number of patients with different levels of visual impairment according to the WHO classification n(%)**

Variable	BCVA range	n (236)	Consanguineous marriage (62)
Mild to moderate visual impairment	20/60<BCVA≤20/200	122 (51.6)	40 (32.8) <sup>a</sup>
Severe visual impairment	20/200<BCVA≤20/400	84 (35.6)	16 (19)
Blindness	BCVA<20/400	30 (12.7)	6 (20)

BCVA: Best-corrected visual acuity. <sup>a</sup>P<0.05.

**Table 3 Distribution of cases categorized by age and the causes of visual impairment n(%)**

Age (a)	Choroid and retina	Optic nerve	Nistagmus	Congenital cataract	Glaucoma	Total
<15	12	6	14 (41.1)	-	-	34
15-30	42 (50)	6	28	-	2	84
31-50	28 (56)	6	14	2	-	50
>50	66 (97)	-	-	-	2	68
Total	148	18	56	2	4	236

**Table 4 Distribution of choroidal and retinal causes of visual impairment by age**

Age (a)	Macular dystrophy	Albinism	Retinitis pigmentosa	Stargart disease	DRP	AMD	ROP	Other	Total
<15	-	6	-	2	-	-	4	-	12
15-30	10	8	2	14	-	-	-	8	42
31-50	6	6	4	10	2	-	-	-	28
>50	2	-	6	2	14	38	-	4	66
Total	18	20	12	28	16	38	4	12	148

DRP: Diabetic retinopathy; AMD: Age related macular degeneration; ROP: Retinopathy of prematurity; Other: Degenerative myopia, coroidal coloboma, tamoxifen toxicite, trauma.

**Table 5 Frequency distribution of causes of visual impairment by parental consanguineous marriage n(%)**

Consanguineous marriage	Nistagmus	Stargart, macular dystrophy	Optic nerve	ROP	Retinitis pigmentosa	AMD	DRP	Other	Total
Positive	20 (32.3)	18 (29)	8 (12.9)	2 (3.1%)	8 (12.9)	-	-	6	62
Negative	38 (21.8)	26 (14.9)	10 (5.7)	2 (1.1%)	4 (2.3)	38	16	40	174
Total	56	46	18	4	13	38	16	45	236
P		<0.05			<0.001				

ROP: Retinopathy of prematurity; AMD: Age related macular degeneration; DRP: Diabetic retinopathy; Other: Degenerative myopia, coroidal coloboma, tamoxifen toxicite, trauma.

All patients had some degree of visual impairments. BCVA less than 20/400 (blindness) was seen in 12.7% of the patients. Parental consanguinity was significantly higher in the group of mild to moderate visual impairment (32.8%) ( $P<0.05$ ), Table 2 displays the number of patients in each level of the visual impairment.

The main causes of low vision were categorized based on age. The most common cause (62.7%) of visual impairment in all ages exact under 15 years old group was retinal and choroidal diseases. Nistagmus was the most common cause under 15 years old group (Table 3).

We also examined the most common disorders in the retinal subgroup by age and we found that the distribution of the retinal disorders varied by age. The most common retinal disorder in the patients between 15-50 years old was macular dystroph. Above 50 years of age, it was degeneration of the macula (16.1%) and diabetic retinopathy 16 (6.7%), respectively Table 4.

In the retinal and choroidal group, age related macular degeneration were seen in 38 patients (16.1%) and diabetic retinopathy, retinitis pigmentosa, Stargart disease and macular dystrophy were seen in 16 (6.7%), 12 (5.1%), 28 (11.9%), and 18 (7.6%) patients, respectively (Table 4). Parental consanguineous marriage was evaluated according to the cause of the disease. With retinitis pigmentosa ( $P<0.001$ ), stargart and macular dystrophy ( $P<0.05$ ) consanguinity rate was significantly higher. Age related macular degeneration (AMD) and Diabetic retinopathy (DRP) group, there was no consanguinity that was statically significant (Table 5). Eighty-eight (37.3%) patients Galilean type; 116 (49.2%) patients keplerian type telescopic glasses recommended. Eighteen (7.6%) patients had not improvement in visual acuity with LVA. Forteen patients (5.9%) were approved the magnifier for near vision. In most patients, the use of LVAs improved both near and distance visual function (Table 6).

**Table 6 Vision changes with Low Vision Aids (LVA)**

Variable	<sup>1</sup> mean BCVA	Median near vision
Without LVA	0.14 (±0.11)	No;5
With LVA	0.14 (±0.11)	No;2

BCVA: Best-corrected visual acuity; <sup>1</sup>P=0.001.

## DISCUSSION

This study demonstrated that the leading causes of visual impairment, based on the major headings of The International Classification of Diseases (ICD - 10) classification, in patients in Turkey were retinal and choroidal diseases and nistagmus. Optic nerve and optic tract diseases, congenital cataract and glaucoma were less common causes. Although this pattern of prevalence was observed across all age subgroups, the main causes responsible for visual impairment, according to the subheadings of the classification, varied among them. In patients less than 15 years of age, nistagmus and albinism were the most prevalent. In adults between 15 to 50 years of age, stargart, macular dystrophy and retinitis pigmentosa were the most prevalence and in those above 50 years of age, degeneration of the macula and diabetic retinopathy were the most common causes.

In a retrospective study conducted with 4711 patients who presented to a rehabilitation clinic in Tubingen (Germany), age-related macular degeneration was the most important cause (40%) of visual impairment, followed by tapetoretinal dystrophy, optic nerve atrophy and diabetic retinopathy<sup>[15]</sup>. In another retrospective study with 573 patients in a Malaysian rehabilitation clinic, the most common cause of visual impairment varied by age; congenital disease was most prevalent in individuals less than 30 years of age, those between 30 and 60 years of age reported the most retinitis pigmentosa, and those greater than 60 years of age reported the age-related macular degeneration<sup>[16]</sup>. In a study conducted with 362 students in three schools for the blind in Tehran (Iran), the rate of severe low vision was 8.9%. The most common cause of low vision in that study was retinal diseases (51%). Cataract, optic nerve atrophy, corneal and anterior segment disease, glaucoma, anophthalmia and globe malformations were other common causes of low vision in schools in Tehran<sup>[17]</sup>. In the current study, 41.8% of our patients were above 50 years of age, which might explain the high prevalence of age-related macular degeneration and diabetic retinopathy. This finding is in line with the global data on visual impairment in the year 2002<sup>[3]</sup>.

The extant literature thus far suggests a preponderance of males with visual impairments. Nguyen *et al*<sup>[15]</sup> found that 58.9% of the patients were male. In the current study, the majority (66.1%) of the enrolled populations were male as well. Male gender were higher in all age groups In this sample. It appears that more man than women seek services from the rehabilitation clinic.

Parental consanguinity was present in 62 (26.3%) patients, most commonly between the ages of 15-30 were significantly higher in the group (50%) ( $P < 0.001$ ). However, most

causes of low vision in this age group, stargart, macular dystrophy, nystagmus, respectively. Of genetics in the etiology of these diseases are known to be effective. For this reason, consanguinity is still a big problem in Turkey. About consanguinity and regarding the transmission of these diseases, training of people seem to be essential. The two most common diseases among patients older than 50 years old in our study were age-related macular degeneration and diabetic retinopathy. Parental consanguinity is very rare over 50 years old (5.9%). This result was also statically significant. There was no parental consanguinity in both age related macular degeneration and diabetic retinopathy patients groups. Less of an impact in the etiology of these diseases are genetic.

In the current study, retinal and choroidal diseases were the most common causes of visual impairment. There are three explanations possible: 1) retinal and choroidal diseases are less treatable and reversible compared with other ophthalmologic problems, 2) retinal and choroidal diseases are more frequent in our society because of parental consanguinity and 3) patients with these types of ophthalmic problems may be referred more often to rehabilitation clinics by ophthalmologists. Cataract problems are the main cause of visual impairment in most population-based studies<sup>[3]</sup>. Other causes in decreasing order of frequency include refractive errors, trachoma, glaucoma, macular degeneration and retinal diseases<sup>[11]</sup>. Since our study was conducted with referred patients to a visual rehabilitation clinic, the two common diseases (*i. e.* cataract and refractive errors), were not seen as major causes of vision loss in our population. These findings confirm the first hypothesis because patients with cataract and refractive errors are usually managed properly in our society.

The assumption of a higher prevalence of retinal diseases among our society. It is possible that not only the acquired but also the congenital forms of retinal disease are highly prevalent in our population. This finding converges with similar studies conducted in Brazil<sup>[18]</sup>, Germany<sup>[15]</sup>, India<sup>[19]</sup>, Thailand<sup>[20]</sup>, and the Netherlands<sup>[21]</sup> that show that the most common cause of low vision is retinal diseases. Conversely, in studies performed in Ethiopia<sup>[22]</sup> and Uganda<sup>[23]</sup>, the anterior segment of diseases had a more important role in vision loss, which might reflect a high prevalence of infectious diseases and vitamin A deficiencies in these societies. Further, in a study with 3210 children in Brazil, the most common cause of visual impairment in children with multiple disabilities were optic nerve atrophy (37.7%) and cortical blindness (19.7%). The authors emphasized the physical examination of other parts of the body in a child with optic nerve atrophy<sup>[18]</sup>. In the current study, optic nerve diseases had the second highest prevalence after the choroidal and retinal diseases.

The kind of prescribed LVAs are mostly dependent upon visual function of each patient, not the causes of low vision. So each visual function should be considered first when prescribing the LVAs<sup>[24-26]</sup>.

In most patients, both near and distance BCVA were much improved when using the LVAs. This suggests that LVAs are effective in the improvement of visual function<sup>[27-29]</sup>.

In summary, although young patients still represent the majority of patients visiting the low vision clinics, we found that the number of elderly low vision patients is increasing and macular degeneration is becoming a leading cause of low vision. Rehabilitation using LVAs has improved the visual function of low vision patients.

According to research, consanguineous marriage rate, 25% in İstanbul, 20%–30% in other cities. In our study results were similar. Parental consanguineous marriage rate was 26.3% at our patients. Also in our country, such as sickle cell anemia and Thalassemia carrier has a high frequency of genetic diseases. Genetic diseases, is an important problem for Turkey, there is no cure, the maintenance costs are too high and causing the psychological trauma.

This study did not involve all-blind people because they are not usually referred to rehabilitation centers. It should be cautioned that because this current study only examined those who presented at a rehabilitation clinic, the generalizability of these results is limited. However, this study identified the irreversible and untreatable causes of low vision in our society and could be helpful in planning for screening programs.

Unfortunately consanguineous marriage is a big problem still in Turkey. About consanguineous marriage and regarding the transmission of these diseases, training of people seem to be essential. Moreover, the results of these types of studies can be used in creating and informing guidelines of practice for rehabilitation clinics. Further studies especially in other cities with larger populations are warranted.

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