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# Al-Shifa Journal of Ophthalmology

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- **Editorial: Growing Menace of Diabetic Retinopathy in Pakistan**
- **Knowledge And Attitude of Parents Towards Children with Strabismus**
- **Outcome with Spectacles and Telescopes among Albino Patients**
- **Association of Consanguinity with Keratoconus**
- **Knowledge about Diabetic Retinopathy**
- **Protection from COVID-19 Hazards**
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# **Al-Shifa Journal of Ophthalmology**

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Al-Shifa Trust Eye Hospital, Rawalpindi**

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## Growing Menace of Diabetic Retinopathy in Pakistan

Ume Sughra

According to the World Health Organization Diabetes mellitus (DM) affects about 400 million adults globally, a number that is anticipated to be doubled by 2030.<sup>1,2</sup> The International Diabetes Federation (IDF) estimated that the global population with diabetes mellitus (DM) to be 463 million in 2019 and 700 million in 2045.<sup>3</sup> Diabetic retinopathy, a microvascular disease is a common complication of diabetes, defined as severe non-proliferative diabetic retinopathy (NPDR) or proliferative diabetic retinopathy (PDR) or the presence of diabetic macular edema (DME) caused by high blood sugar levels damaging the back of the eye (retina).<sup>4</sup> One third of diabetic patients suffered from diabetic retinopathy, mostly non-proliferative diabetic retinopathy. Diabetic retinopathy increased markedly after the age of 60 years, which could be due to the longer duration of diabetes.<sup>5</sup> It damages the blood vessels of the light-sensitive tissue at the back of the eye (retina) that results in blindness if left undiagnosed and untreated. It is estimated that 20 years after diagnosis, those with type 1 diabetes and 60% of those with type 2 diabetes will have some level of retinopathy. It is the main source of visual impairment in patients aged 20–74 years and approximately 4 million individuals worldwide are estimated to be losing their sight from diabetic retinopathy.

The risk of development and progression of diabetic retinopathy is closely associated with the type 1 diabetes, longer duration of diabetes, advancing age, poor glycemic control, high blood pressure and elevated serum lipids.<sup>6-10</sup>

The burden of diabetic retinopathy is expected to remain high through 2045 worldwide, disproportionately affecting countries in the Middle East and North Africa and the Western Pacific. The prevalence of Diabetic retinopathy was highest in Africa (35.90%) and North American and the Caribbean (33.30%) and was lowest in South and Central America (13.37%).<sup>11</sup> These estimates are expected to rise further due to the increasing prevalence of diabetes, ageing of the population and increasing life expectancy of those with diabetes.

The prevalence of diabetic retinopathy in Pakistan was found to be 28.78% in all diabetics and that of vision threatening diabetic retinopathy was 28.2% of all diabetic retinopathy and 8.6% of all diabetics. There is lack of conclusive data to highlight the problem of diabetes and DR to generate enough advocacy for the policy makers to plan a “National program” to address diabetes related blindness. Hakeem R et al reported a prevalence of diabetes as 7.6% to 11%. In a recent press release by Baqai Institute of Diabetology and Endocrinology (BIDE), prevalence of diabetes in Pakistan is 26%.<sup>12</sup>

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# Knowledge And Attitude of Parents Towards Children Suffering from Strabismus Presenting in a Tertiary Care Eye Hospital at Rawalpindi

Naima Shakeel<sup>1</sup>, Ayesha Babar Kawish<sup>2</sup>, Momina Javed<sup>1</sup>, Mussarat Ali<sup>1</sup>, Fareeha Ayyub<sup>1</sup>, Sadaf Qayyum<sup>1</sup>

## Abstract

**Background:** Early diagnosis and treatment is very crucial for successful treatment, which is only possible if the parents notice their child's strabismus early and take timely interventions

**Objectives:** The purpose of this study was to assess knowledge and attitude of parents toward children suffering from strabismus coming to Al-Shifa Eye Hospital, and to find out the association between parental knowledge and psychosocial aspects of strabismus.

**Material and methods:** A cross-sectional study was conducted from October 2019 to December 2019 at orthoptics department of Al-Shifa Trust Eye Hospital, Rawalpindi. The study population included all the parents of children with strabismus presenting in orthoptics department of Al-Shifa Trust Eye Hospital, Rawalpindi, fulfilling the inclusion criteria. Sample size calculated was 75 parents. Non-probability convenient sampling technique was used. Data was collected through interview-based questionnaire. Data analysis was done using Statistical Package for Social Sciences (SPSS) Version 20. Chi square test was used to find association between outcome variable and independent variable.

**Results:** The results showed that the association between education level of parents and their knowledge about strabismus is not statistically significant. Further the association between education level of parents and their attitude towards their child's strabismus is not statistically significant however, there is significant association between knowledge and attitude. 72.1 % (N=31) of parents with good attitude had good knowledge.

**Conclusion:** The success of strabismus treatment primarily depends on the age of presentation when early diagnosis and treatment is possible. Lack of knowledge about strabismus delays the entire process of identification and treatment of strabismus. *Al-Shifa Journal of Ophthalmology* 2021; 17(1): 8-12. © Al-Shifa Trust Eye Hospital, Rawalpindi, Pakistan.

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## Introduction:

Strabismus is a condition in which both eyes are not properly aligned and point in opposite direction. There are mainly six muscles in each eye which are responsible for eye movements. If one eye has weaker muscles than the other eye, it will often move in other direction instead of focusing at the desired object. If this condition persists, the weaker eye can become permanently misaligned leading to a condition known as amblyopia, or 'lazy eye.'

(1)



Prevalence, according to the research that had been done in areas including America, Singapore, Iran and India, the estimated pooled prevalence (95% CI) of any strabismus, exotropia, and esotropia was 1.93%, 1.23%, and 0.77%.<sup>(3)</sup> This study revealed that 1 in every 50 people had strabismus, which severely affects their quality of life.<sup>(3)</sup> Overall estimated prevalence of strabismus in Pakistan is 5.4%. Out of this, 2.5% strabismus patients are under the age of 5 years while 2.9% patients are over the age of 5 years. The national prevalence of 5.4% suggests that there are 7.02 million patients with strabismus in population of 130 million.<sup>(3)</sup>

Strabismus is one of the most common ocular disorder affecting children, with a prevalence of 3 to 5% among children.<sup>(2)</sup> Early diagnosis and treatment is very crucial for successful treatment, which is only possible if the parents notice their child's strabismus early and take timely interventions.<sup>(2)</sup> Other complications associated with strabismus include diplopia, asthenopia, decreased vision, poor development of binocular single vision, stereopsis, and cosmetic blemish.<sup>(2)</sup> Besides this the affected children may develop psychological problems such as poor self-esteem and self-confidence, difficulty in adjustment and making new friends and other behavioral & psychosocial problems.<sup>(2)</sup> The success of treatment for strabismus mainly depends upon on the age of presentation, therefore its early diagnosis and treatment is very important.<sup>(2)</sup> The treatment options include use of corrective spectacles, prisms, orthoptic exercises, amblyopia therapy and surgery.<sup>(2)</sup>

If the eyes can no longer maintain their correct position or focus on the same object, the result is problems with binocular vision. In cases of strabismus, or cross-eye, the eyes are not properly aligned, with one is drifting from its normal position. Strabismus causes the brain to have difficulty synthesizing visual images from

each eye, resulting in impaired binocular vision and depth perception. Amblyopia, or lazy eye, is another condition that causes binocular vision problems. Amblyopia occurs when the brain ignores input from one eye, affecting depth perception and other visual abilities. Strabismus is the commonest cause of amblyopia that can be prevented or treated if detected early.<sup>(4)</sup>

### **Materials and Methods:**

A cross-sectional study was conducted from October 2019 to December 2019 at orthoptics department of Al-Shifa Trust Eye Hospital, Rawalpindi. Study was carried out in Orthoptics department of Al-Shifa Trust Eye Hospital (ASTEH), Rawalpindi, Pakistan. The study population included all the parents of children with strabismus presenting in orthoptics department of Al-Shifa Trust Eye Hospital, Rawalpindi, fulfilling the inclusion criteria. Sample size calculated was 75 parents. Non-probability convenient sampling technique was used. The inclusion criteria of our study were children of age between 1 to 14 years and parents of children having strabismus while both genders were included. The exclusion criteria of our study were children with any mental/physical disorder, children with strabismus with positive family history, unresponsive individuals / parents were excluded and non-volunteers. Data was collected through interview-based questionnaire which were typed in English. All data was collected by principal author herself. questionnaires were validated. Patients were evaluated under the supervision of senior optometrist. Parents of children up to 14 years of age participated in this study.

Data was coded daily and entered into SPSS. Questionnaires were protected in file that was only in excess of researcher. SPSS files and backup were password protected. After entering data in SPSS, the data was cleaned by running frequencies in SPSS. Knowledge and attitude were outcome

variables. Age, gender, area of residence, socio-economic status, occupation of parent, parental education level, Psycho-social impact were independent variables. Data analysis was done using Statistical Package for Social Sciences (SPSS) Version 20. Confidence level taken was 95%. Descriptive analysis was generated both for dependent and independent variable. Categorical data was presented in the form of percentages and frequencies and quantitative data was presented in the form of mean, median and standard deviation.

Chi square test was used to find association between outcome variable and independent variable. The test was applied on all applicable independent variables and outcome variables, a significance level of 5% was used for all inferential statistics.

Research was conducted after approval of Institutional Review Board of Pakistan Institute of Ophthalmology. Permission was taken from the supervisor and head of department of orthoptics department. Informed consent was taken from every Participant/parent of children presenting in Al-Shifa Eye Hospital, Orthoptics department. Information of every individual kept confidential.

### **Results:**

A total of 75 participants were included in the study. Among them majority of patients were males i.e., 54.7% (N=41) and remaining 45.3% (N=34) were females. Their mean age was  $16.15 \pm 4.116$  with minimum 1 year and maximum 14 years. Most of the patients arrived at OPD from urban area i.e., 57.3% (N=43) and remaining 42.7% (32) from rural. Among total number of participants 12% (N=9) were illiterate, 20% (15) were under matric, 21.3% (N=16) were matric, 16% (N=12) were undergraduate, 8% (N=6) were graduate and 21.3% (N=16) were post graduate.

Out of 75 participants 26.7% (N=20) had the knowledge about strabismus and 73.3% (N=55) did not know about strabismus. In majority of cases the strabismus was detected by parents i.e., 73.3 (N=55) and remaining was detected by teachers 1.3% (N=1), primary eye care physician 10.7% (N=8) and others 14.7% (N=11). Most of the parents i.e., 93.3% (N=70) noticed strabismus themselves. 38(50.7%) parents felt that strabismus is affecting the vision of their child. 30(40%) and 11(14.7%) parents noticed that their child is facing difficulty in reading and depth perception respectively. 39(52%) parents had the knowledge about the treatment of strabismus and 36(48%) parents were not aware of treatment of strabismus.

Out of 75 participants 77.6% (N=59) parents were concerned about their child's condition and 64.5%(N=49) complaint that their child's cosmetic appearance is being affected by squint. 75%(N=57) parents felt that their child will face difficulties in future due to strabismus. 60.5%(N=46) said that they feel uncomfortable when someone asks about their child's strabismus. The results showed that the association between education level of parents and their knowledge about strabismus is not statistically significant. The results showed that the association between education level of parents and their attitude towards their child's strabismus is not statistically significant.

A relationship between the knowledge of parents and their attitude towards their child's strabismus had been found using chi square which shows that there is significant association between knowledge and attitude. 72.1 % (N=31) of parents with good attitude had good knowledge.

## Discussion:

The study was carried out to assess the knowledge and attitude of parents toward children suffering from strabismus presenting in ASTEH. In majority of the cases, strabismus is a treatable condition that requires identification and treatment at early age. However, whether the treatment is given in a timely manner depends on parents' knowledge and attitude. Lack of knowledge and information among parents adversely affects the age of presentation and management of strabismus. Parents in this study were segregated into six groups, namely, illiterate, under-matric, matric, under graduate, graduate and post graduate. Among total participants 12% were illiterate, 20% were under matric, 21.3% were matric, 16% were undergraduate, 8% were graduate and 21.3% were post graduate.

The study shows that there is no significant association between education level and knowledge or attitude of parents. 56% of parents had good knowledge and 58.1% of parents had good attitude. Study also shows that there is significant association between knowledge and attitude. 72.1% of parents with good attitude had good knowledge. A study on knowledge and attitude towards strabismus among parents of Saudi children with strabismus showed similar results. <sup>(5)</sup>

The study further showing knowledge and attitude of parents tells us that 26.7% parents had the knowledge about strabismus and 73.3% did not know about strabismus. While the study conducted on knowledge, attitude and practice towards strabismus in Cheha District, Central Ethiopia showed that 73.6% had the knowledge about strabismus and 26.4% had less knowledge. <sup>(5)</sup>

In majority of cases the strabismus was detected by parents i.e., 73.3. 50.7% parents felt that strabismus is affecting the vision of their child. 40% and 14.7% parents noticed that their child is facing difficulty in reading and depth perception respectively.

77.6% parents were bothered by their child's strabismus and 64.5% complaint that their child's cosmetic appearance is being affected by squint. 71.1% parents felt that strabismus will hinder their child's performance at school. 75% parents felt that their child will have few opportunities in future due to strabismus. 60.5% said that they feel uncomfortable when someone asks about their child's strabismus. 25% parents used to feel that their child will have difficulty in making friends. 21.3% parents noticed psychological changes due to strabismus in their children. 52% parents had the knowledge about the treatment of strabismus. 48% parents were unaware of the possible treatments for the condition hence did not seek proper medical advice and timely treatment. This led to the delayed presentation of patients, resulting in higher susceptibility to strabismic amblyopia. A study on knowledge, attitude and practices among parents towards children with strabismus in Goa concluded the similar results. <sup>(6)</sup>

The study also shows that most of parents were also under constant stress because of the views expressed by others about their child's condition and the sense of discomfort experienced when they were asked about their child's "abnormal" eyes. Parents' knowledge regarding strabismus is very crucial to prevent strabismic amblyopia since once amblyopia sets in, it leads to a loss of vision and loss of binocularity, reducing of vision later, and thus, hampering job opportunities, Children suffering from strabismus is not only cosmetic concern but also it had more implications. It has adverse effects on their self-esteem, self-confidence, ability to obtain work, and interpersonal relationship. These children face problems at school and work which severely impairs their socialization, symptoms of which are lower aspiration level and remoteness from social groups - both primary and secondary. <sup>(7)</sup>

### **Conclusion:**

The success of strabismus treatment primarily depends on the age of presentation when early diagnosis and treatment is possible. This is only possible with better awareness about strabismus among parents. Lack of knowledge about strabismus delays the entire process of identification and treatment of strabismus. Hence health education is very important to increase the awareness and change myths about strabismus; thus, making early diagnosis and successful treatment possible. Early diagnosis and treatment will also help prevent psychosocial problems and behavioral disorders among children affected with strabismus and improve academic performance and prevent the development of amblyopia.

### **Recommendations:**

There is need of awareness programs in general public for strabismus and importance of its treatment. Strabismic amblyopia is avoidable (preventable or treatable) condition. Early diagnosis and prompt treatment of strabismus by regular screening or checkup would reduce the risk of strabismic amblyopia which may cause unnecessary visual impairment so that every individual becomes independent and capable of achieving life goals.

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# Types of Refractive Errors and Visual Outcome with Spectacles and Telescopes among Albino Patients

Mutahir Shah<sup>1</sup>, Saifullah<sup>2</sup>, Maryam Firdous<sup>2</sup>, Farah Amin<sup>2</sup>, Sufian Ali Khan<sup>1</sup>

## Abstract

**Purpose:** To determine types of refractive error and Visual Outcome with spectacles and telescopes in Albinism

**Methodology:** This was a Cross Sectional study conducted in Low Vision Department of Shifa Eye Foundation Hospital Haripur. Study duration was 6 months from March 2020 to September 2020. Sampling technique was non-probability convenience sampling. Refractive errors were evaluated subjectively and objectively. Best corrected visual acuity with spectacles and telescopes were assessed binocularly.

**Results:** A total of 37 patients with Albinism were examined and divided into two groups i.e., Ocular Albinism (OA) and Oculocutaneous Albinism (OCA). The frequency of OCA was 29 and that of OA was 07. OCA Astigmatism was the most common refractive error about 72.41%. Astigmatism was found with a mean value of 2.71D. Myopia had a mean value of -6.7D, while the degree of Hyperopia was found with a mean value of 7.20D. Main Refractive Errors found among OA was Myopia and Astigmatism with equal percentage of 42.85%, Hyperopia was 14.28%. Myopia was found with a mean value of - 9.66D. Astigmatism was found with a mean value of 3.5D and Hyperopia was 5.12D. In OCA the uncorrected Visual Acuity (VA) was found with a mean value of 0.99 Log MAR. In OA it was found with a mean value of 0.90 Log MAR. With Spectacle in OCA it was 0.82 Log MAR while with Galilean telescope the value was 0.54 Log MAR. With Keplerian telescope it was 0.25 Log MAR and in OA with Spectacles the mean value was 0.72 Log MAR.

**Conclusion:** Astigmatism was the most significant refractive error among Albino patients. VA had improved with spectacles and telescopes. Keplerian telescope improved vision more significantly compared with Spectacles and Galilean telescope. *Al-Shifa Journal of Ophthalmology 2021; 17(1):13-17. © Al-Shifa Trust Eye Hospital, Rawalpindi, Pakistan.*

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## Introduction:

The term albinism is applied to a group of genetically determined disorders of melanin deficiency in vital part of the body especially skin and eyes<sup>1</sup>. Albinism occurs with an overall frequency of 1 in 18,000 in the United States<sup>2</sup>. It is a heterogeneous group of conditions which are clinically divided into Oculocutaneous Albinism (OCA) and Ocular Albinism (OA). In OCA the skin, hair and eyes are involved while in OA only the ocular features are present. Reduced or absent tyrosinase activity due to mutant alleles of the tyrosinase genes is a frequent cause of Oculocutaneous Albinism<sup>3</sup>. All forms of albinism are

characterized by Nystagmus Photophobia and reduced Visual Acuity. Different study showed that OCA is a group of 4 autosomal recessive disorders and OA is X-linked but uncommonly autosomal recessive.

The major sign and features present among the all type of albino patients are nystagmus usually pendular, high refractive error of various types, light sensitivity, iris Trans-illumination from decreased pigmentation, deficiency of pigments in retina especially peripheral to the posterior pole, reduced central acuity usually ranges from 20/25 to 20/200<sup>4</sup>. Nystagmus usually begins from the age of 2-3 months of life<sup>5</sup>. The severity of visual acuity depends upon the amplitude of nystagmus and degree of pigmentation.

As albinism is a hereditary condition so no medical and surgical treatment are available. The treatment options available for albino patients are mostly optical<sup>6</sup>. Optical treatment options consist of Spectacles, Tinted lenses for photophobia Hat and low vision aids<sup>4</sup>. Most of the low vision aids used for albino patients consist of telescopes which may be of Galilean or Keplerian types<sup>7</sup>. Galilean type of telescopes has minus ocular lens and plus objective lens and vice versa for Keplerian<sup>8</sup>. The distance between the ocular lens and the exit pupil is referred to as eye relief<sup>9</sup>.

The purpose of this study was to find the best possible optical treatment option available for albino patients that will enhance the visual outcome like visual acuity.

### **Subjects and Methods:**

It was a Cross Sectional Study on 37 albino patients presenting in low vision department of Shifa Eye Foundation Hospital Haripur. The sampling technique was non-probability convenience sampling. Total duration of the study was 6 months. All patients with Albinism were included. Both OA and OCA with subjects having

age from 5-30 years including both genders were included. Any type of deviation i.e., esodeviation or exodeviation and media opacities were excluded. The study was conducted after approval had been accorded by the hospital ethical committee. As per the protocol of the hospital, every patient referred to the low vision clinic had his complete ophthalmologic examination done in the general OPD. Any patient having signs of albinism was referred to low vision clinic. For optical and non-optical treatment, the patient then lands in the low vision clinic. The patients were selected for the study by convenient sampling taking into consideration the inclusion and exclusion criteria.

All the patients of albinism were assessed for visual acuity measurement. Visual acuity was taken on ETDRS test chart. Subjective refraction of the patient was done. Near vision was taken with light house NV chart or with the help of light house NV chart. The visual acuity was recorded in terms of log Mar. Then different types of LV aids i.e., telescopes were applied depending upon the need of the patient and severity of the disease. Telescopes used for distance vision correction included Galilean and Keplerian telescopes. Visual acuity was checked with these telescopes and the results were documented. All the observation along with demographic information of patients was noted on a pre-designed structured Proforma. Data analysis was done using Statistical Package for Social Sciences (SPSS) version 25. Descriptive statistics like frequencies and standard deviation was presented for types of refractive error, Gender and visual outcome with spectacles and different optical aids. Mean and standard deviation was calculated for numerical variable i.e., Visual acuity uncorrected and Visual acuity corrected with spectacles, telescopes and for types of refractive errors.

**Results:**

Total 37 patients of Albinism were included in the study divided into two types, Ocular Albinism (OA) and Oculocutaneous Albinism (OCA). The number of subjects for OCA was 29 and that for OA was 8.

Result of the study showed that in Oculocutaneous Albinism Astigmatism was the most common refractive error about 72.41%. Degree of Astigmatism was ranging between (-4.50- +4.50) with a mean value of 2.71D. The nature of astigmatism was with the rule. Myopia and Hypermetropia showed equal representation with 13.7%. Degree of myopia ranges between (-2.0-15.75D) with a mean value of -6.7D, while the degree of Hyperopia ranges between (+3.50-+12.00) with a mean value of +7.20D.

Main Refractive Errors found in Ocular Albinism subjects were Myopia and Astigmatism with equal percentage of 42.85%, Hypermetropia was 14.28%. Degree of Myopia was ranging between (-2.0-16.0) with a mean value of 9.66D. Degree of Astigmatism was ranging

between (-6.5-+4.0) with a mean value of 3.5D. Degree of Hypermetropia was ranging (2.25-8.0) with a mean value of 5.12D.

In OCA the uncorrected Visual Acuity was ranging between (1.6-0.6 Log MAR) with a mean value of 0.99 Log MAR. In OA it was ranging between (1.2-0.5 Log MAR) with a mean value of 0.90 Log MAR. With Spectacle in OCA, it was ranging between (1.2-0.5 Log MAR) with a mean value of 0.82 Log MAR (0.21 SD). With Galilean Telescope the range was (0.78-0.1 Log MAR) with a mean value of 0.54 Log MAR (0.18 SD). With Keplerian Telescope the range was (0.6-0.0 Log MAR) with a mean value of 0.25 Log MAR (0.18 SD).

In Ocular Albinism with Spectacles the range was (1.2-0.22 Log MAR) with a mean value of 0.72 Log MAR and a standard deviation of 0.16. With Galilean Telescope it was ranging between (0.8-0.2 Log MAR) with a mean value of 0.49 Log MAR (0.18 137.SD). With Keplerian telescope it was ranging between (0.6-0.0 Log MAR) with a mean value of 0.24 Log MAR (0.18 SD)

Table-1: Distribution based on types of Albinism

Type Of Albinism	Frequency	Percentage
Oculocutaneous Albinism	29	80.55%
Ocular Albinism	08	19.45%
Total	36	100

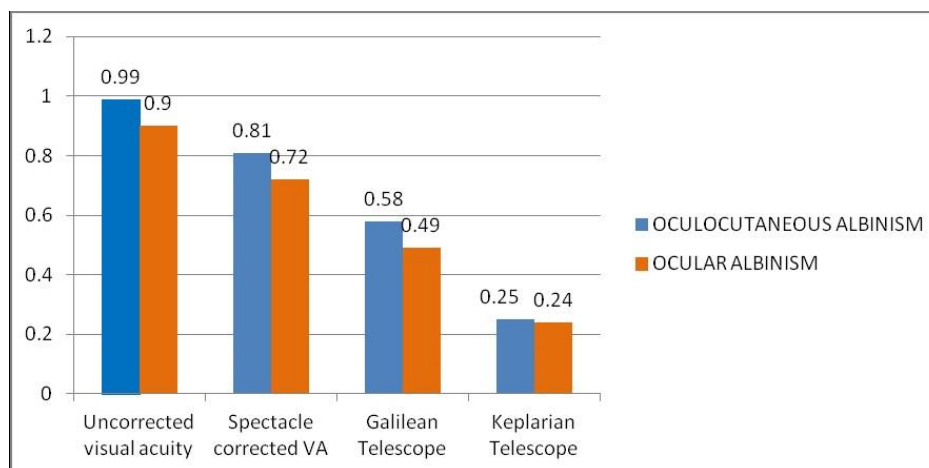


Fig-1 shows graphical representation of uncorrected VA in oculocutaneous albinism and ocular albinism

### Discussion:

This study showed that majority of patients visited LVA department of were mainly males. This did not mean that male were more prone to Albinism as it is a genetic disorder and both gender are affected equally it was due to social values that restrict females to reach to a hospital.

The percentage of oculocutaneous albinism was 80.55% and of ocular albinism was 19.45%. It showed that oculocutaneous albinism was more common in this region.

The results from showed different types of refractive errors among both types of albinism. According to our study the types of refractive error found among oculocutaneous albinism was mainly astigmatism. The frequency of oculocutaneous albinism with Astigmatism was found to be the most common refractive error about 72.41%. Degree of Astigmatism was ranging between (-4.50-+4.50) with a mean value of 2.71D. The nature of astigmatism was with the rule. A similar study was published in a journal<sup>10</sup> showed that the average astigmatism was found 2.37D and with the rule in nature<sup>11</sup>. The results of our study regarding magnitude of refractive errors was similar with nominal difference with the above stated study. Myopia and Hypermetropia were found equal with a frequency of 4

patients and 13.7%. Degree of myopia ranges between (-2.0-15.75D) with a mean value of -6.7D. Hypermetropia was 13.7%. Degree of Hyperopia ranges between (+3.50-+12.00) with a mean value of +7.20D.

Another study reported that myopia and Hyperopia in Albinism was ranging from (-9.78 to +8.88D)<sup>11</sup>. The study also showed that ocular albinism represented with varying degree of refractive errors. The frequency of astigmatism and myopia were same followed by hypermetropia. Out of 07 patients with ocular albinism 03 had astigmatism, 03 had myopia and only 01 patient had hypermetropia. Degree of Myopia was ranging between (-2.0-16.0) with a mean value of 9.66D, Degree of Astigmatism was ranging between (-6.5-+4.0) with a mean value of 3.5D and Degree of Hypermetropia was ranging between (2.25-8.0) with a mean value of 5.12D. A similar study had published in a journal showing that myopia and Hyperopia in Albinism was ranging from (-9.78-+8.88D)<sup>7</sup>. The result of our study showed similarity with the above study.

The current study showed that the uncorrected VA in OCA was 0.99 LogMAR and 0.90 Log MAR in OA. Study found that visual acuity was improving with spectacles and low vision aids. The mean



corrected VA with spectacles was 0.82 (6/38) with 0.21 SD. The mean corrected VA with Galilean telescope was 0.54 (6/19) (0.18 SD) and with Keplerian VA was 0.25 (6/9.5) (0.18 SD). A similar study published in a journal showed that a large proportion of these albinos had noticeable refractive errors (astigmatism and high myopia) and Poor visual acuity usually less than 6/30 (20/100) with spectacle correction<sup>8</sup>.

### **CONCLUSION:**

Astigmatism was the most significant refractive error in Albinism. Visual Acuity and functional vision improved with spectacles, filters and telescopes. Compared with Spectacles and Galilean telescope Keplerian telescope improve vision more significantly, but price, lighter weight and easy use make Galilean more useful in Albinism.

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## Association Of Consanguinity with Keratoconus Among Patients Presenting at A Tertiary Eye Care Hospital, Rawalpindi; Case Control Study

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### Abstract:

#### Objective:

The aim of this study was to find out an association between keratoconus with consanguinity of the parents of the patients and also find out the other risk factors associated with keratoconus.

**Methodology:** This case-control study was conducted over the duration of four months from October 2019 to January 2020 and it included patients who were diagnosed with keratoconus at the cornea department of Al-Shifa Trust Eye Hospital, Rawalpindi (ASTEH). The control group included age and sex matched individuals who were randomly selected from patients presenting in general OPD of ASTEH without keratoconus. Study subjects were asked to fill structured questionnaire that included demographic and geographic details, questions on ocular and general health and keratoconus profile history. Data was analyzed by using SPSS version 20. A Chi-square test was used to check the association of consanguinity with keratoconus.

**Results:** 115 cases and 230 controls were included in this study among them 52.2% were male in case groups and 48.6% were female in the control group. Majority of the respondents presented with positive ocular allergy in both cases 78.8% and controls 52.4%. There is significant association between the consanguinity and keratoconus with the p value of 0.00.

**Conclusion:** This study concluded that eye rubbing, ocular allergy, consanguinity and having family members with keratoconus emerged as significant risk factors for KC. *Al-Shifa Journal of Ophthalmology 2021; 17(1):18-23. © Al-Shifa Trust Eye Hospital, Rawalpindi, Pakistan.*

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### Introduction:

Keratoconus (KC) is non inflammatory progressive thinning and steeping of the paracentral part of the cornea involve one or both eyes at the same time and also known as corneal ectasia.<sup>1</sup> The Patient with keratoconus initially feels the gradually reduction of vision due to myopia or irregular astigmatism.<sup>2</sup> The etiology of the diseases is related to both genetic as well as environmental factors such as ocular allergy, continuous rubbing of eye ball, atopy and UV exposure to the eyes.<sup>3</sup> Consanguinity is cousin marriage or the marriage between the relatives is associated with infancy morality, congenital defects such as deafness, blood diseases, heart diseases, Tuberculosis, facial clefts and mental and physical handicap.<sup>4</sup> Previous

studies showed that there are high chances of keratoconus among relative marriages.<sup>5,6,7</sup> In Pakistan prevalence of consanguinity is 50 to 80% but there is little evidence on the systematic evaluation of keratoconus with consanguinity according to best of our knowledge. Therefore, this was aimed to find the association between the consanguinity and keratoconus and also find the other risk factors associated with the keratoconus.

### **Materials and Methods:**

This study was conducted at cornea department of Al-Shifa trust eye hospital, Rawalpindi. This comparative case-control study included those patients diagnosed with keratoconus both unilateral and bilateral were selected as a case group and for controls age and sex matched individuals having no keratoconus were selected as a control group. Patients with any other ocular disease except keratoconus and not willing to participate were excluded from this study. Moreover, for control groups patients with keratoconus were excluded.

By using the systematic random sampling technique, data was collected from both groups over the periods of five months from October 2019 February 2020. This was done to ensure inclusion of all participants fulfilling the study criteria during the study duration, as it will allow a more accurate analysis of the association of keratoconus with consanguinity.

A structured questionnaire was used for data collection, the first part of the questionnaire collected information about the independent variables like socio-demographics of the patients while the second part included questions about general medical history, ocular history, and keratoconus history. The questionnaire was written in the English language initially, but it was translated into the local language to ensure comprehension of the participants. Permission was taken from the Ethical

review committee of Al- Shifa trust eye hospital before data collection. Moreover, verbal informed consent was also taken from every individual before they participated in this study. Face and content validity was checked by circulating it to experts in the field. All the data was collected by the primary researcher itself. Data was collected from those who were diagnosed with keratoconus after the detailed ophthalmic investigations. Confidentiality of the patient's data was maintained and ethical values of research were properly considered and followed at every step of the study.

The responses in the questionnaire were recorded and analyzed using Statistical package for social sciences (SPSS) version 20. The descriptive analysis was done on the categorical and continuous variables. Percentages and frequencies were reported for categorical variables while for mean and standard deviations well as minimum and the maximum value was reported for continuous variables. The inferential analysis was done by applying Chi-Square of independence to find the association between dependent and independent variable.

### **Result:**

A total of 340 participants were included in this study. The cases 33.2% (N= 113) and controls 66.8% (N=227) were in 1:2 proportion. The number of males who participated in this study was 52.2% (N=59) in cases 51.8% (N=176) and in controls 51.5% (N=117). Mean age of the respondents was 20.12 with (SD ±6.64) ranging from 9-44 years. A proportion of 35 (10.3%) participants belonged from rural areas and 305 were 89.7% (N=305) with 23.2% (N=79) belonging from working class and 76.8% (N=261. 80% (N= 272) respondents were unmarried Table No: 1.

Majority of the respondents presented with positive ocular allergy in both cases (N=89, 78.8%) and controls (N=119, 52.4%). Most

of the respondents included in the study have history of first cousin marriages in both cases 67.3% and controls 30%) while unrelated marriages history in cases (N=18,

15.9%) and controls (N=96, 42.3%). Table No 2.

Chi-Square test was performed to find the association between dependent and independent variables. Table 3.

Table 1: Demographics Characteristics

Variables	Categories	Cases	Controls	Total Participants
Gender	Male	59(52.2%)	117(51.5%)	176(51.8%)
	Female	54(47.8%)	110(48.5%)	164(48.2%)
Residence	Rural	10 (8.8%)	25 (11%)	35(10.3%)
	Urban	103 (91.2%)	202 (89.0%)	305(89.7%)
Working Status	Working	25 (22.1%)	54 (23.8%)	79(23.2%)
	Non-working	88 (77.9%)	173 (76.2%)	261 (76.8%)
Marital Status	Unmarried	94(83.2%)	178 (78.4%)	272(80%)
	Married	19(16.8%)	49 (21.6%)	68(20%)
Family History		22(15%)	16, (4.4%).	

Table No: 2 General Medical & Ocular History

Variables	Categories	Cases N (%)	Controls N (%)	Total Participants
Ocular Allergy	Positive	89 (78.8%)	119 (52.4%)	206 (60.6%)
	Negative	24 (21.2%)	108 (47.6%)	132 (38.8%)
Atopy	Positive	4 (3.5%)	6 (2.6%)	9(2.6%)
	Negative	109 (96.5%)	221 (97.4%)	329 (98.8%)
Asthma	Positive	1 (0.9%)	14 (6.2%)	14 (4.1%)
	Negative	112 (99.1%)	213 (93.8%)	324 (95.3%)
Diabetes Mellitus	Positive	2 (1.8%)	11 (94.8%)	12 (3.5%)
	Negative	111 (98.2%)	216 (95.2%)	326 (95.9%)
Smoking	Positive	9 (1.8%)	28 (12.3%)	36 (10.6%)
	Negative	103 (91.2%)	197 (86.8%)	299 (87.9%)
Consanguinity	Double first cousin	9 (8%)	12 (5.3%)	21 (6.2%)
	First cousin	76 (67.3%)	68 (30%)	144 (42.4%)
	Second cousin	7 (6.2%)	22 (9.7%)	27(7.9%)
	Cousin once-removed	3 (2.7%)	29(12.8%)	32(9.4%)
	Unrelated marriage	18 (15.9%)	96(42.3%)	114 (33.5%)
<b>Ocular History</b>				
Spectacle	Positive	86(76.1%)	105(46.3%)	190 (55.9%)
	Negative	27(23.9%)	122(53.7%)	148 (4%)
RGP History	Positive	4 (3.5%)	8 (3.5%)	12 (3.5%)
	Negative	109 (96.5%)	219 (96.5%)	326 (95.9%)
Permanent use of Glasses	Positive	20(17.7%)	41 (18.1%)	61 (17.9%)
	Negative	93(82.3%)	186 (81.9%)	277 (81.5%)
Eye rubbing	Positive	89(78.8%)	131 (57.7%)	219 (64.4%)
	Negative	24(21.2%)	96 (42.3%)	119 (35%)
Previous use of Topical Medication	Positive	85 (75.2%)	77 (33.9%)	162 (47.6%)
	Negative	28 (24.8%)	150 (66.1%)	176 (51.8%)
Ocular Surgery	Positive	36(31.9%)	29 (12.8%)	65 (19.1%)
	Negative	74(65.5%)	198 (87.2%)	270 (79.4%)

Table No 3: Association between Dependent & Independent Variables

Independent variable	Categories	Cases N %	Controls N %	x <sup>2</sup> (df)	p value
Gender	Male	59	117	0.014(1)	0.907
	Female	54	110		
Ocular Allergy	Positive	89	119	22.03(1)	0.000
	Negative	24	108		
Atopy	Positive	4	6	0.212(1)	0.645
	Negative	109	221		
Asthma	Positive	1	14	4.992(1)	0.025
	Negative	112	213		
Diabetes mellitus	Positive	2	111	1.941(1)	0.164
	Negative	111	216		
Smoking	Positive	9	28	1.487(2)	0.475
	Negative	103	197		
Spectacle History	Positive	86	105	1.487(2)	0.475
	Negative	27	122		
RGP History	Positive	4	8	0.000(1)	0.994
	Negative	109	219		
Consanguinity	Double first cousins	9	12	50.581(4)	0.000
	First cousins	76	68		
	Second cousins	7	22		
	Cousins once-removed	3	29		
	Un-related marriages	18	96		
Family history of Keratoconus	Positive	22	16	17.576 (6)	0.007
	Negative	90	211		

**Discussion:**

A total of 340 participants were enrolled in this study with 1:2 ratios of cases 33.2% and controls (66.8%). Our study showed that the percentage of parental first-cousin marriage was 67.3% among patients with keratoconus (KC) and 30% among patients without keratoconus these findings are supported by one of a study in Israel which showed 59.3% were first cousin marriage among cases and 30.0% among control group. The family history of keratoconus among cases was 16% whereas among controls were 4.4% in this study.<sup>8</sup> While in study of collaborative longitudinal evaluation of keratoconus observed the rate

of family history of 13.5% and study in Israel reported 21.7% family history with keratoconus.<sup>9, 10</sup> The present study showed significant association between consanguinity and keratoconus which provide the strong support of genetic contribution to KC, moreover several studies suggested a number of genes which played an important role in pathogenesis of KC including *VSX1*, *SOD1*, *COL4A3*, and *COL4*.<sup>11</sup> Gordon-Shaag et al also support our study results and demonstrated a strong link between KC and parental first-cousin consanguinity.<sup>8</sup> Parental cousin marriages causes the increased risk of ocular diseases such as infantile strabismus, strabismus

with convergence excess, Esotropia with anisometropic amblyopia, Duane syndrome and monocular elevation deficiency.<sup>12</sup> Consanguinity is more common among Asia and Africa where almost 50% marriages are cousin marriage and among them almost one third of all marriages are first cousin marriage.<sup>13</sup> Same trend was observed in Tehran where 69% were first cousins marriage among total consanguinity.<sup>14</sup> In Pakistan approximately 60% were consanguinity among them 80% were first cousin marriages.<sup>15</sup> Keeping in mind this trend of cousin marriages many new couples need to understand the consequences of consanguinity for their offspring. Therefore, public health practitioner should counsel the public and screening guidelines should be adapted to evaluate the consanguineous couples and their offspring. In present study there is strong association between KC and ocular allergy because it is main risk of increasing the severity of keratoconus.

The association between KC and atopy was not significant whereas asthma is strongly associated with KC. Smoking has been shown to be significantly less common in KC patients than in controls and this study confirms non-significance between cases and controls. Association with diabetes mellitus was also non-significant with p value >0.05 in both cases and controls. The main limitation of our study was that it included only referred patients of KC who were came for their management of KC, outdoor patients were not included in our study. In short, cousin marriages have an influence in the pathogenesis of keratoconus so we should conduct the health awareness seminar and educate the peoples about the effects of cousin marriages on the health of their offspring's.

### **Conclusion:**

This data provides strong evidence of a genetic predisposition to KC as evidenced by consanguinity. It would further seem that a genetic background may not be

sufficient to lead to ocular morbidity and interaction with an environmental risk factor such as eye rubbing or allergy may be necessary in the causation of the disease. The association of consanguinity with Keratoconus stresses the need for public health awareness of the harmful consequences of inbreeding.

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# Assessment of Knowledge About Diabetic Retinopathy and Motivating Factors for Screening Among Diabetic Patients In Rawalpindi

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## Abstract:

**Background:** Diabetic Retinopathy (DR) is the fifth leading cause of blindness in worldwide and fourth cause of blindness in Pakistan. This study assesses level of knowledge about DR among diabetic patient and the factors which motivate patient for DR screening.

**Methodology:** A cross-sectional study was conducted (including 370 diabetic patients) from October 2019 to December 2019 at three general hospitals of Rawalpindi. A structured questionnaire was used to collect the information from patients it consisted of three sections i.e., demographic profile of diabetic patient, knowledge about DR, the factors that motivates patient for DR screening. After collection of data, it was coded and entered into SPSS version 20. Then the data were analyzed using Chi-square test.

**Results:** About 45.7% had high knowledge about DR and 42.4% had high motivation level for DR screening. Socio-economic status like education level, family history of DM, and monthly income had influence on knowledge of DR and shows statistically significant effect on knowledge about DR (p-value < 0.001). Education level also shows statistically significant effect on the factors that motivate patients for DR screening (p-value < 0.001).

**Conclusion:** The study showed lack of awareness of DR among diabetic patients in the study population. There is an urgent need for early DR screening and for refining knowledge and awareness of diabetes and DR in Rawalpindi, Pakistan. *Al-Shifa Journal of Ophthalmology 2021; 17(1):24-29. © Al-Shifa Trust Eye Hospital, Rawalpindi, Pakistan.*

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## Introduction:

Diabetes mellitus (DM) is the major health problem of 21<sup>st</sup> century which is associated with macro-vascular and micro-vascular diseases such as kidney diseases, neural damages, cerebrovascular diseases and eye complications<sup>1</sup>. There are 280 million diabetic patients universally which is assumed to be twofold by 2025<sup>2</sup> due to population growth, aging, physical inactivity and obesity<sup>3</sup>. Globally, individuals who living with diabetes among them 50% are undiagnosed and when they diagnosed, many individuals have already developed complications from their condition<sup>4</sup>. The prevalence of this disease in Pakistan was 12% according to Diabetes National Survey in 2010<sup>2</sup>. Pakistan is expected to become the 4th foremost country in the number of patients with DM,



in 2020<sup>5</sup>. The complications related to DM are awful. It is expected worldwide that about 15000 to 39000 individuals lose their vision because of DM and about 14.6% developed diabetic retinopathy (DR) after 5 years duration of DM at the age of 40 and above.<sup>6</sup> The most common complication of DM is diabetic eye disease (DED) containing of numerous complaints like diabetic macular edema (DME), diabetic retinopathy, cataract and glaucoma. If not diagnosed on time and treated properly can lead to visual impairment and total vision lost<sup>4</sup>. The most common micro-vascular disease is diabetic retinopathy (DR) and it is the most devastating complication occurs in approximately one third of the patient with diabetes<sup>7,8</sup>.

DR is the major cause of blindness worldwide<sup>9</sup>, it causes damage to the walls of small blood vessels of retina which leak or become blocked due to prolonged high blood glucose level and unstable the structure and function of the retina<sup>4,10</sup>. Individuals with diabetes need to be well-informed about and involve in self-care to control the risk factors including long term diabetes, deprived glycemic control dyslipidemia, nephropathy, pregnancy, and gender, and poorly controlled hypertension<sup>7,11, 12</sup>. DR contributes 4.8% of the international causes for impaired vision<sup>13</sup>. DR is linked with duration of diabetes in diabetic patients and occurs in both type 1 and type 2 DM<sup>(11, 14)</sup>. Control of the adaptable risk factors through intermittent eye checkups and appropriate interventions has been shown to delay the advancement of DR<sup>11</sup>. In many clinical strategies regular DR screening is suggested but in practice only 58% of all DM patient were screened<sup>15</sup>.

Barrier have been described by some studies which include: patients rely on that they do not have DR, feel embarrassment about poor glycemic control, unease about the treatment, contradictory priorities, believing that other hospital eye

appointments or regular optometrist appointments test for DR and lack of awareness of the importance of Screening<sup>16</sup>. Awareness of DR and its consequences for vision is an important requirement for attendance for screening<sup>17</sup>.

### **Subjects and Methods:**

A hospital based cross-sectional study was conducted to assess the knowledge of DR and screening motivation factors among diabetic patients. Data was collected over duration of three months (October 2019 to December 2019) at three general Hospitals of Rawalpindi (Holy Family hospital, Military Hospital and Benazir Bhutto Hospital). All patients with diabetes mellitus (DM) presenting in the above mentioned three general Hospitals (endocrinology department) of Rawalpindi were included in this study. Data was directly collected from the patients because the questionnaire based on the knowledge or awareness of patients regarding diabetic retinopathy (DR). Sample size of 370 was calculated This was calculated with 60% awareness about Diabetic Retinopathy (DR) among diabetic patient (M. Saleh Memon, 2014) by using open-Epi. The technique for this study was Convenient sampling. In all of the general hospitals of Rawalpindi only three hospitals were selected randomly. Inclusion criteria was Diabetic patient were included in the study. Age criteria was from 20 to 70 years. Both genders were included. Individuals who were not willingly participating were excluded from the study. Patients with any physical/mental disabilities were also not the part of the study. Data was collected from participants through a structured questionnaire. It consists of three sections: Demographic profile of diabetic patient, Knowledge about DR. The factors that motivate patient for DR screening the cross-sectional study was conducted on almost 370 patients who had diabetes mellitus and who visiting the 3 general hospitals of Rawalpindi (Military Hospital, Holy Family hospital and Benazir Bhutto

Hospital). After taking informed consent from the individuals then they were interviewed with the help of structured questionnaire.

The study investigated two outcome variables: Knowledge and the factors that motivates patient for DR screening. The two outcome variables were evaluated with the help of questionnaire based on different questions related to knowledge of DR and motivation factors for DR screening. Independent variables were socioeconomic demographics which included: Gender, age, education level, marital status, residence, occupation, monthly income, Type of DM, Duration of DM, Family history of DM, Control of DM. After collection of data, it was coded and entered into Statistical Package for Social Sciences (SPSS) software version 20 on daily basis and saved thereafter. Before starting of analysis data errors were omitted by reevaluating the questionnaire. After that, data transformations were carried out and continues variables (age) was transformed into different categories. Then data analysis was done in two phases, descriptive analysis followed by inferential statistics.

Descriptive statistics were generated for all variables. Categorical data was presented in the form of frequencies and percentages. Mean, standard deviations, and ranges were reported for continuous variables. Median was reported for heterogeneous continuous variables with high standard deviations. Chi-square test for independence was used for finding association between outcome variable and independent variable. The test was applied on all the applicable independent variables and outcome variable. A significance level of 5% was used for all inferential statistics. The study was conducted after approval by Institutional Review Board. Permission was also taken from the medical superintendent of all the three General Hospitals where the study was conducted.

Verbal informed consent was obtained from each and every patient included in the study. It was insured that data collected was used only for academic purpose and confidentiality of the data was ensured.

### **Results:**

A total 370 individuals were interviewed with the help of structured questionnaire and all of them were agreed to participate. Response rate was 99%. The mean age of individuals having diabetes mellitus was 50.41 years (SD=10.89) ranging from 25 to 70 years. Almost half (48.4%) of the patient were female. 27.8% respondents were illiterate and 25.9% had higher education of university level. More than half (80.5%) of the total respondents were married. Majority of the patients (264, 71.4%) belonged to Rawalpindi.

About 150 (40.5%) respondents had type 2 diabetes mellitus and 139 (37.6%) didn't knew the type of DM. More than one half (250, 67.6%) of the patient had family history of diabetes mellitus (DM). Half of the total patients were unemployed currently (214, 57.8%) and almost half (190, 51.4%) of the patients had monthly income above PKR 30000/-. About 35.1% (130) patients knew that DM effect eyes while 31.4% (116) didn't knew about the damage caused by DM to body.

The study participants (370) with a mean diabetic duration of 9.10 years (SD=7.12) ranging from 1 year to 32 years. Almost all of the patients were having treatment for diabetes. More than half 79.7% (295) respondents knew that diabetes had effects on eyes. Less than half (136, 36.8%) had knowledge about diabetic retinopathy (DR). 30.0% of the respondent gained knowledge about DR from their diabetologist and 105 (28.4%) patients were informed by ophthalmologist. Mostly of them knew that DR cause blindness only 8.6% (32 of the respondent) told that DR could not cause blindness. As DR can be progressed if not manage properly but only

50.5% knew that poor control of Diabetes Mellitus caused the progression of DR. Laser surgery, the one of the most common treatment option available for DR but only 27% (100 of the respondent) had knowledge about this treatment option. And 30% didn't know about any treatment option available for it.

Only 24.3% had knowledge about the yearly eye examination for a diabetic patient. 40.5% (150) respond that there was no need for regular eye checkup if diabetes was under good control. More than half (67.8%) of the patients knew that if DR was not treated on time it can lead to avoidable blindness. 80.3% of the individuals had knowledge that a good control of DM can prevents eye problems.

Mostly (56.8%) individuals were up to date on their given appointment date. 47.8% of the respondents were agree to went for regular eye checkup as their eye doctor told them even if they don't have any problem in their eye. Personal screening attitude of 46.8% were positive. High percent 48.1% (178) of individuals were disagreed with the sentence that social media plays its role for DR screening and motivate them to do so. 53.0% of the patients were encouraged by the family for DR screening. 38.9% were disagreed that the physician encouraged them for DR screening. 53.0% (196) were agreed that the cost of DR screening was very high that's why they were not really motivated for screening.

Association of demographic variables with knowledge regarding diabetic retinopathy shows significant results with education level family history of diabetes and monthly income i.e., p value less than (0.001). Association of demographic variables with motivation factors for DR screening also showed highly significant results with education level and duration of diabetes.

### **Discussion:**

In the present study findings suggest that 45.7% of the respondents had high knowledge about diabetic retinopathy and about 79.7% knew that eyes could be affected by diabetes but only 36.8% were aware of diabetic retinopathy as ocular complication of diabetes. This result is similar to the other study conducted in South India by Nithin Keshav Srinivasan et al<sup>1</sup> which shows that 71.9% had knowledge that diabetes could damage eyes and only 17.01% were aware of diabetic retinopathy that it could be caused by diabetes. More than half (54.3%) of the patients were aware that DR cause blindness.

One study by Lui and Chen showed that only 36.6% of subjects with Diabetes were aware of DR as a diabetic complication that could result in blindness.<sup>18</sup> In the present study 30.0% of the individuals get information from their physician about diabetes related eye complication and 28.4% were informed by ophthalmologist. The study done in KSA showed 67.2% of the patients had obtained information about diabetes-related eye complications from healthcare workers, while 18% of the patients had attained this information from media.

In this study 30.0% didn't knew about any of the treatment option available for diabetic retinopathy but about 37.9% of the individuals had knowledge about laser and eye injections that were available to treat DR. A study done in Jeddah, Saudi Arabia by Sami H. Alzahrani presented that more than 34% of subjects said that surgery and laser are not available treatment options for DR<sup>11</sup>. Mostly, the studies done on DR were KAP studies, but this study also includes the factors which motivates patients for DR screening. High motivation level for diabetic retinopathy screening was 42.4%. 56.8% of the individuals were generally up to date on their required screening tests. Mostly (48.1%) of the respondents were not motivated from social media and they

disagreed with the statement that social media play its part in motivating individuals for DR screening. 53.0% of the respondents were encouraged for screening by their family members because 67.6% had family history of diabetes and about 48.1% were motivated from their family history. 53.0% of the individuals agreed that the treatment cost for DR was the barrier against their motivation level. This was also studied by Sneha Lingam a BSc student in India and conducted that glucose and blood pressure screening was a main motivator for (17.7%) DR screening<sup>18</sup>. Possibly, care providers' lack of skill and training or their extreme focus on treatment rather than prevention might be the reason for low awareness about DR, but this desires additional exploration<sup>11</sup>. Ophthalmologists, general physicians and health care workers have to working in union to aware diabetic individuals regarding eye complications cause by DM and should educate about the treatment option available for it and to motivate the individuals for screening tests. Along with awareness, self-help and life style should also be improved<sup>19, 20</sup>. Early detection by screening and timely treatment for DR are effective in preventing blindness<sup>15</sup>

### **Conclusion:**

This study showed lack of awareness of DR among diabetic patients in the study population. Considering the association of DR with DM, the growing prevalence of DR may be a potential burden on community and health systems. There is an urgent need for early DR screening and for refining knowledge and awareness of diabetes and DR in Rawalpindi, Pakistan.

Shortage of knowledge concerning the need for screening for diabetic retinopathy was found to be a major barrier to compliance with regular screening. Health messages conveyed through mass media should be simple, original, use different formats, and be repeated many times.

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# Protecting Healthcare Workers from COVID-19 Hazards

Shehnilla Shujaat<sup>1</sup>, Khalid Iqbal Talpur<sup>1</sup>

## Abstract

**Objective:** To ensure, the quality and proper management of ophthalmic patients & protection of health care providers amidst hard time of COVID-19.

**Study design:** Study design is observational study.

**Place and duration of study:** The place of our study was Sindh Institute of Ophthalmology and Visual Sciences and duration of this study was nine months from 1<sup>st</sup> April 2020 to 31<sup>st</sup> December 2020.

**Methods:** Ophthalmic patients and ophthalmologists were at higher risk of being infected by the novel corona virus because of close proximity during examination and procedures, so special strategies were adopted to protect them. Hospital as the hub of ophthalmic procedures was at the top target for this disease. The patients were ensured to be provided special care of hand wash / sanitizer & face mask for protection against covid-19. All the SOPs were strictly followed by health care providers, that included use of protective equipment like face masks, hand gloves, hand sanitizers. Also, special campaign was carried out, which educated people about complete awareness of symptoms pertaining to coronavirus. Elective procedures were deferred to prevent the spread of virus and emergency patients were attended on urgent basis.

**Results:** Gradual and timely change in strategies helped to ensure continued ophthalmic care in the challenging time of COVID-19 without interruption. Simultaneously the safety of health care providers was also being taken care of.

**Conclusion:** Precautionary steps to protect the health of ophthalmic practitioners may be considered during hard times and crisis like that of pandemic covid-19. *Al-Shifa Journal of Ophthalmology 2021; 17(1):30-35. © Al-Shifa Trust Eye Hospital, Rawalpindi, Pakistan.*

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## Introduction:

In December 2019, in Wuhan, China, a number of pneumonia patients were admitted with unknown causes. On 31<sup>st</sup> December, it was traced to be caused by a novel corona virus. It was named COVID-19 (corona virus disease 2019) later renamed as SARS-CoV 2 (severe acute respiratory syndrome coronavirus 2). It was first reported as epidemic by an ophthalmologist, Li Wenliang who contracted Corona Virus while treating a glaucoma patient with conjunctivitis<sup>1</sup>. The disease then spread very rapidly to the other countries of the World and WHO declared it as a global pandemic in March 2020<sup>2</sup>.

There is much speculations about its origin. The recent data shows that the bat CoV and the human 2019-nCoV share a recent

common ancestor<sup>3</sup>. The primary source of interpersonal transmission is via direct contact or through droplets due to coughing and sneezing of an infected person<sup>4,5</sup>. The virus also remains viable on inanimate objects such as metals, plastic and glasses for up to 9 days<sup>6</sup>.

The incubation period ranges from 2 to 14 days and infectivity remains 1 to 2 days before onset of symptoms to the end with the resolution of symptoms<sup>1</sup>. The inhaled virus SARS-CoV-2 likely binds to epithelial cells and starts replicating. ACE 2 (angiotensin converting enzyme receptor type 2) is the main receptor for SARS CoV-2.<sup>5,7,8</sup> The kidney, intestine and conjunctiva also have ACE 2 receptors but in small numbers.

Ophthalmologists are at high risk of getting the 2019-nCoV due to close proximity with the patients during examinations, investigations and procedures. The COVID-19 patients may present with conjunctivitis as its first manifestation. The viruses have found in some specimens of tears and conjunctival secretions of COVID-19 patients. When the aerosol droplets from infected person come in contact with conjunctiva, it gets infected and some are drained in the nasopharynx and trachea through nasolacrimal pathway<sup>9</sup>. The ophthalmic instruments may also become transmitter of infections to the patients and the health professionals. An asymptomatic COVID-19 patient may encounter in the hospital and can cause cross infection in the area.

Until now, prevention is a key factor in breaking transmission and to reduce the chance of getting the disease. Every patient presented with conjunctivitis should be enquired of fever, cough and respiratory distress to rule out coronavirus disease. Whenever possible, keep a distance of 1 meter with patient. Ask patient to avoid touching and keep silence during slit lamp examinations. Frequent hand washing with

soap and water for 20 seconds and with chlorhexidine alcohol hand rub after each ophthalmic examinations and therapeutic procedures is highly recommended to prevent cross infections. The use of mask is mandatory both for patients and the ophthalmologists. He should use respirators like N95 during high-risk patient examinations like conjunctivitis with respiratory symptoms and operative procedures. Gloves should be used during handling of body secretions, fluid and lesions and periorbital and orbital manipulations. The contact lens wearers are encouraged to use glasses to reduce touching the eyes. It has been accepted that 2019-nCoV is sensitive to ultraviolet irradiation and heating<sup>10</sup> so it is important to sterilize the instruments with heating and lipid solvents after each use. If it is possible, the non-urgent ophthalmic examinations and operations should be postponed. The ophthalmologist should not forget to change outdoor clothes and taking shower soon after return to home from workplace and to keep the clothes at separate place. The doctor should measure his own temperature at least twice daily and if having fever, a diagnostic PCR test should be performed<sup>1</sup>.

Conjunctivitis may be the first manifestation of 2019 nCoV [11]. A patient with COVID-19 may shed virus in his tears<sup>12</sup>. An ophthalmologist can make himself safe and his patients by strictly following the preventive measures. He also has a role in confronting and notifying the disease and warns the patients about patient related responsibilities<sup>1</sup>. Patient literacy rate in Pakistan is very low, so majority of people were not following the precautions regarding corona virus seriously and they were initially visiting hospitals without taking care of SOPs. As a results hospital administration had to take special measures to implement the stringent safety policies in the light of recommendations given by World Health Organization.

**Materials and Methods:**

All the patients visiting to OPD were provided with water and hand wash for cleaning of their hands. then directed to cover their nose and mouth with a mask.

Patients visiting to Out Patient Department were asked questions related to COVID 19 symptoms like cough, fever, sore throat, dyspnea and red eye. Patients having simple conjunctivitis were checked with torch and provided with drops. Patients who needed the urgent ophthalmic care were attended with slit lamp examination and indirect ophthalmoscopy. These mostly included patients presenting with pain due to infections, trauma, ulcers, acute congestive glaucoma and patients with sudden loss of vision.

Patients who were given dates for elective procedures, like phacoemulsification, lens matter aspiration and IOL implantation, ptosis surgeries were deferred till the crisis diminishes. Follow up patients were

provided with eye drops advised for longer follow ups.

Emergency procedures were attended as soon as possible. Those procedures included corneal lacerations, corneal perforations, acute angle closure glaucoma, lens induced glaucoma, dropped nucleus during cataract surgery, and dropped intraocular lens, postoperative endophthalmitis and fresh Rhegmatogenous Retinal Detachment. All these procedures were attended with SOPs.

**Results:**

Gradual and timely change in strategies helped to ensure continued ophthalmic care during the challenging time of COVID-19 without interruption. Simultaneously the safety of health care providers was also being taken care of. Table 1, 2 and 3 show the reduced number of patients entertained in outpatient department, laser and operation theatre departments.

Table No. 1: Maximum Per Day Data Comparison Before COVID-19 & After COVID-19

S. No.	No. of Patients Before COVID-19	No. of Patients After COVID-19
1	800	35

Table No. 2: Per day data of Laser procedures before COVID-19 & after COVID-19

S. No.	Name of Procedures	No. of patients Before COVID-19	Age (Years)	No. of patients After COVID-19	Age (Years)
1	YAG Laser	18	55-60	01	55-60
2	PRP Laser	09	40-50	01	40-50



Table No. 3: Comparison of Elective and Emergency surgical procedures before COVID-19 &amp; after COVID-19

S. No.	Name of Procedure	No. of patients Before COVID-19	Age Range	No. of patients After COVID-19	Age Range
1	Phaco with IOL	17	45-60	06	45-60y
2	ECCE with IOL	04	50-65	02	50-65y
3	LMA + IOL	02	05 y/o	01	8 y/o
4	Repair	01	11 y/o	01	07 y/o
5	EUA Cyclo Fundus	02	07-11	01	07 y/o
6	Lensectomy	03	01-06		
7	Phaco + Trab	01	19 y/o		
8	PPV with Endolaser	04	18-55		
9	PPV with SO / Gas	01	70 y/o		
10	Secondary IOL	01	50 y/o		
11	Squint Surgery	01	07 y/o		
12	DCR with Tube	02	19-60		
13	Conjunctivital Flap	01	66 y/o		
14	Evisceration	01	36 y/o		

### Discussion:

It was observed that ratio of patients dropped significantly in all areas of eye hospital services. Number of patients in outpatient department, Operation Theatre and Lasers (YAG and PRP) affected because of covid-19 intensity, so that in our hospital. With little bit difference, Ateev Mehrotra and his colleagues reported that initially in the pandemic the quantity of appointments to ambulatory care practices dropped by almost 60 % <sup>13</sup>.

Average number of outpatient department before covid-19 were 800 per day but significantly reduced to only 35 patients per day in age group of 30 to 40 years. In Operation Theatre, procedures like phacoemulsification and extra capsular cataract extraction were dropped from 21 Surgeries to only 8 surgeries before and after covid-19 pandemic. Another study reported that the initial decrease in visits was most evident in the New England, Mid-Atlantic, and Pacific regions <sup>14</sup>.

Laser Procedures (YAG and PRP) were decreased from 18 (YAG Lasers) and 9

(PRP Lasers) to only 1 (YAG Laser) and 1 (PRP Laser) per day in age group of 30 to 45 years before and after COVID 19 Pandemic. In another study, weekly visits to dermatologists, urologists, and adult primary care physicians, among other specialists, have been exceeding the pre-pandemic baseline. But weekly appointments to certain other specialists, including pulmonologists and behavioral health providers, remain substantially below their baseline<sup>15</sup>.

### **Conclusion:**

Safety of the health professionals including ophthalmologists is very important to fight against the novel corona virus and to reduce the risk of collapse of health system against this lethal virus. Protective modulations should be made in all ophthalmic institutions all over the world to not only bear the present ophthalmic complications but the future crisis as well.

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## Unusual Features of Orbital Xanthogranuloma: A Case Series

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### Abstract

**Purpose:** To describe previously unreported features of patients with orbital xanthogranuloma.

**Methods:** This was a retrospective study of 5 patients with orbital xanthogranuloma treated and followed up within a twelve-year period from January 2007 to December 2018. Their clinical records were reviewed and analyzed.

**Results:** There were 2 cases each of juvenile xanthogranuloma and adult onset xanthogranuloma and one case of Erdheim – Chester disease of the orbit. Three patients were males and 2 were females. Three patients had unilateral disease while in two cases it was bilateral. The main presenting complaints were proptosis and loss of vision seen in 4 cases.

CT scan demonstrated extraconally located fairly defined masses in all except for the case of Erdheim- Chester disease it was bilateral and intraconal. There was bony erosion involving frontal bone in the two patients with adult onset xanthogranuloma. All patients underwent orbitotomy with subtotal excision of yellowish masses from the orbits. They were all followed up for at least one year (range 1 to 12 years). One of the patients with juvenile orbital xanthogranuloma had bilateral recurrence of the lesion after 8 years. Histopathologic examination revealed foamy histiocytes as prominent feature in all cases and variable number of Touton multinucleated giant cells.

**Conclusion:** Orbital xanthogranuloma is a rare heterogeneous group of diseases with similar histopathologic profile. Diagnosis of the disease is made by considering both clinical and histopathological features. Bony destruction and presence of yellowish orbital masses in adult onset xanthogranuloma are being reported for the first time. *Al-Shifa Journal of Ophthalmology* 2021; 17(1):36-45. © Al-Shifa Trust Eye Hospital, Rawalpindi, Pakistan.

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### Introduction

Orbital Xanthogranulomatous Diseases (OXGD) are a group of rare non-Langerhans cell histiocytic disorders of unknown etiology affecting the skin and subcutaneous tissues of the periorbital areas and the ocular adnexa<sup>1</sup>.

Along with local periocular inflammatory lesions, there may be significant systemic features, which may be life threatening<sup>2</sup>. Based on combined clinical and histopathologic profile, four subtypes of the disease are recognized in adults, namely: adult-onset xanthogranuloma (AOX), adult-onset asthma and periocular xanthogranuloma (AAPOX), necrobiotic xanthogranuloma (NBX), and Erdheim-

Chester disease (ECD) <sup>3, 4</sup>. Juvenile xanthogranuloma (JXG) typically affects children <sup>5</sup>. Due to rarity of orbital xanthogranulomas, available treatment modalities are largely empirical with variable outcomes <sup>1, 2</sup>. Probably for the same reason certain possible features of orbital xanthogranulomatous lesion have been largely speculative. For instance, it is postulated that adult onset xanthogranuloma is unlikely to be associated with bone destruction <sup>6</sup>. Here we present our experience in the management of five patients with OXGD and report the presence of orbital bone destruction due to the lesion in two of them with adult onset xanthogranuloma.

### **Materials and Methods:**

This was a retrospective study in which the clinical records of all histopathologically proven cases of orbital xanthogranuloma managed at the Orbit and Oculoplastic unit of a tertiary eye hospital in Pakistan from January 2007 to December 2018 were reviewed. Medical records were analyzed for age at presentation, gender, laterality, symptoms, duration of symptoms, clinical features, radiological features, treatment methods, histopathological diagnosis and outcome. All cases underwent anterior orbitotomy to obtain tissue diagnosis. Institutional ethics committee approval was obtained and the study was conducted in accordance with the Declaration of Helsinki.

### **Results:**

A total of 5 patients were included. Patient demographics, clinical features, treatment results, and follow-up are depicted in Table 1. There were 2 cases each of JXG and AOX while there was one case of bilateral ECD. Three patients were males and two were females. The mean age at presentation was 35 years (range, 3 – 70 years). Three patients had unilateral disease while in 2 cases it was bilateral. The presenting complaints included proptosis (n = 4), loss of vision (n = 4) and

redness of the eyes (n = 1). The mean duration of symptoms before presentation was 1.8 years (range 3 months to 5 years). At presentation, inferior globe displacement was seen in 3 cases (Figure 1). Palpable orbital masses were present in all 5 patients, with one of them experiencing pain in relation to the mass. In the two cases of AOX, the masses were partly cystic on palpation. There was associated loss of vision in 4 of the cases. In two cases loss of vision was due to induced astigmatism and in another two it was due to optic nerve compression as well as exposure keratopathy in the only case of ECD (Figure 2).

The patient with ECD also complained of associated knee and elbow joint pains while one patient with JXG had bilateral flexion deformity of the little fingers (Figure 3). None of the patients was diabetic, hypertensive or asthmatic. There was no neurological symptoms and peripheral lymphadenopathy in any of the cases.

Chest radiograph of the patient with ECD showed streaky densities around the hila and paratracheal suggestive of fibrosis as well as features of osteosclerosis of the knee, ankle and elbow joints (Figure 4). CT scan was done in each case that revealed extraconal fairly defined masses in four cases along with bony erosion involving frontal bone in the two patients with AOX. Patient with ECD showed bilateral intraconal mass lesions abutting both eyeballs with marked proptosis (Figure. 4). Apart from mild leukocytosis and elevated erythrocyte sedimentation rate (58mm/hr.) in the patient with ECD other hematological and biochemical test done on the rest of the patients were within normal limits.

All patients underwent anterior orbitotomy via skin approach with subtotal excision of yellowish masses from the orbits. In the 2 patients with AOX and bony erosion of their orbital roof, the lesion was partly

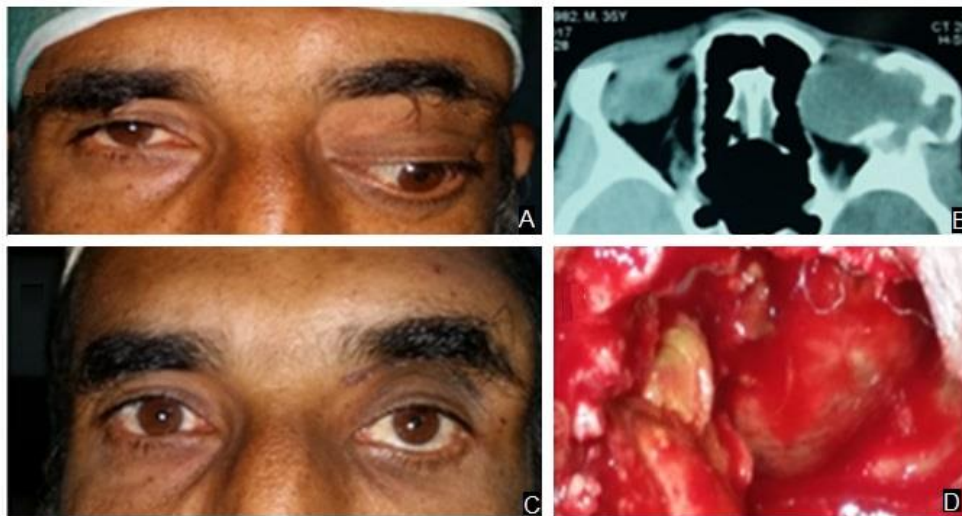
oily/gelatinous and yellowish. The orbitotomy was done in two different surgical sessions for the ECD patient with bilateral disease.

The patient with ECD was started on oral prednisolone 1mg/kg daily prior to surgery and was continued thereafter.

All the patients were followed up for at least one year (range 1 to 12 years). Apart from mild restriction in up gaze, the 2 patients with AOX had almost a near complete resolution of their symptoms. One of the patients with JXG had bilateral recurrence of the mass lesions after 8 years in both inferior orbit/eyelid which were surgically re-excised. Patient has remained stable since the past three and half years now. The patient with ECD developed bilateral severe ptosis after

surgery and the vision remained light perception due to post compressive optic neuropathy and exposure keratopathy. Further systemic evaluation of him for paraproteinemia was negative. He was later lost to follow up after one year.

Histopathologic examination revealed foamy histiocytes as prominent feature in all cases, as were variable number of Touton multinucleated giant cells. A mixed inflammatory cell infiltrate was seen in all the cases as well. Cholesterol clefts were also seen in 1 case of AOX (Figure 5). In the patient with ECD significant fibrosis was seen. Immunohistochemistry done on two of the patients (ECD and one of 2 cases of JXG) showed positivity for S100 protein and CD68 and negativity for CD1a.



*Figure 1. Adult onset xanthogranuloma (A) Patient at presentation showing left inferior globe displacement (B) CT scan showing bone destruction (C) one-year post surgery (D) yellowish fragmented nature of lesion*

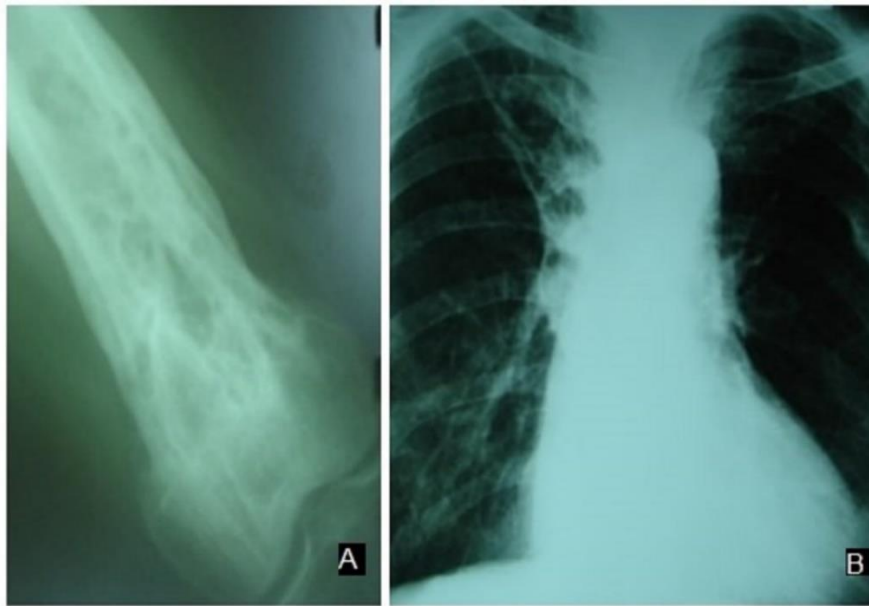


Figure 2: Erdheim Chester disease (A) patient at presentation (B) sagittal and (C) axial CT scan of the orbit showing the lesion occupying most of the orbit (D) yellowish mass removed from the right orbit

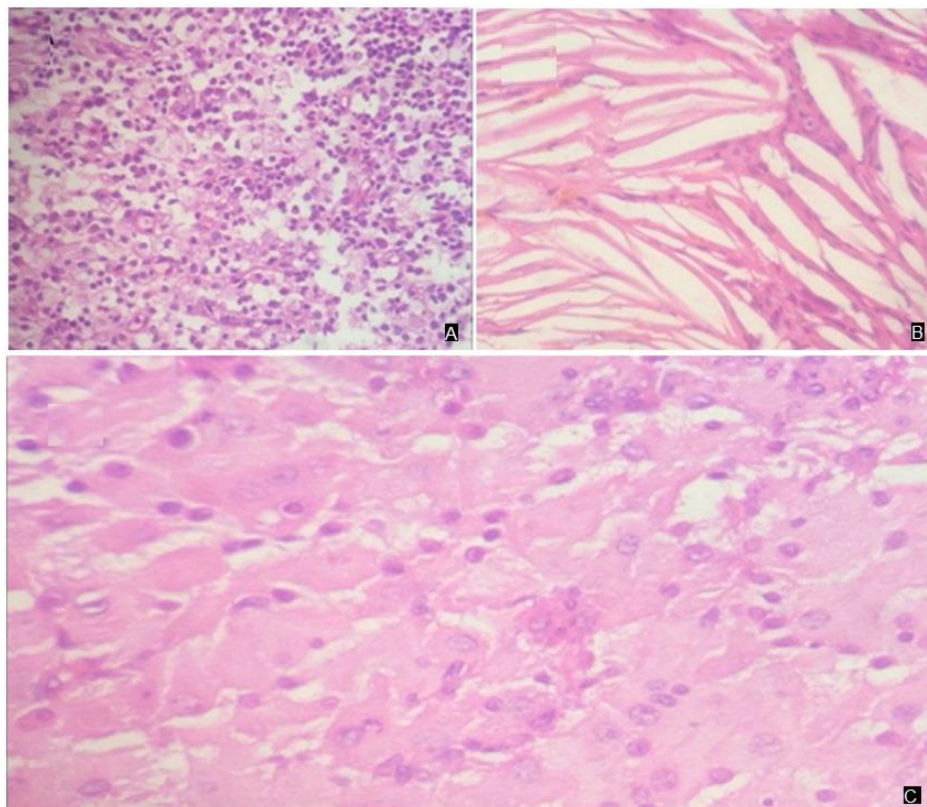


Figure 3: Patient with juvenile xanthogranuloma (A) preoperative photograph at first presentation (B) CT scan showing lesion in the orbits (C) bilateral flexion deformity of little fingers (D) preoperative photograph at recurrence 8 years' after





*Figure 4: X ray (A) femur, showing sclerosis (B) chest, showing hila and paratracheal streaky densities suggestive of fibrosis*



*Figure 5. Hematoxylin and eosin slides show (A) dense lymphohistiocytic proliferation with (B) cholesterol clefts and (C) multinucleated giant cells*



**Table No. I:** Patient demographics, clinical features, treatment and follow-up in 5 patients with orbital xanthogranuloma

No	Sub Type	Later- laity	Clinical features	Systemi c disease	Imaging features	Treatme nt	Prognosis or outcome
1	JXG	UL	Painless proptosis with down displacement of the globe	None	Unilateral circumscribed extraconal superior orbital mass	Excision biopsy	No recurrence
2	JXG	BL	Bilateral multiple swellings in the anterior orbits and periorbita	None	Bilateral circumscribed lesions in the anterior orbits	Excision biopsy	Bilateral recurrence after 8 years. Disease stable after 18 months post re-excision
3	AOX	UL	Painless mass in superior orbit with inferior globe displacement Blurred vision	None	Extraconal superior orbital mass associate with destruction of roof of orbit	Excision biopsy	Stable after 2 years
4	AOX	UL	Painless proptosis	None	Extraconal superior orbital lesion associated with destruction of roof of orbit	Excision biopsy	Stable after one year
5	ECD	BL	Painful bilateral marked proptosis associated with exposure keratopathy and loss of vision	Osteosc lerosis with Knee and elbow joint pains	intraconal mass lesion abutting both eyeballs with marked proptosis. Streaky densities around the hila and paratracheal suggestive of fibrosis. Osteoscleroti s of knee and joint.	Debulk- ing and systemic steroid.	Bilateral post operative severe ptosis. Lost to follow up after one year without recurrence

UL=Unilateral, BL=Bilateral

## Discussion:

The orbital xanthogranulomas share similar clinical and histopathologic characteristics. Clinically superficial lesion impart yellowish coloration of skin and resected lesions are yellowish on gross examination as seen in all our cases<sup>5</sup>. Histologically, the xanthogranulomas is characterized by infiltration of “hallmark cells,” especially foamy histiocytes and Touton-type giant cells, both of which are often negative for S100 and CD1a with varying degrees of fibrosis, necrosis and/or lymphocytic infiltration<sup>6</sup>. Although the orbital xanthogranulomas have overlapping clinicopathological features, some subtle morphological and systemic characteristics of the disease are taken into account in order to adequately subclassify it.

Necrobiotic xanthogranuloma is typically characterized by the presence of subcutaneous skin lesions that tend to ulcerate and become fibrotic and are histopathologically characterized by the presence of necrobiosis surrounded by palisading epithelioid histiocytes, more numerous Touton giant cells and cholesterol clefts. Frequent systemic findings include paraproteinemia and multiple myeloma<sup>4,7</sup>.

ECD, the most devastating of the adult xanthogranulomas, is characterized by dense, progressive, recalcitrant fibrosclerosis of the orbit and internal organs, including the mediastinum, pericardium and pleural, perinephric and retroperitoneal spaces. Orbital involvement is often more posterior and diffuse than with other subtypes and may result in proptosis, ophthalmoplegia, and blindness from optic nerve compression. Bone involvement is common and characteristically presents as diffuse sclerosis of the diaphysis and metaphysis of long bones<sup>7</sup> and death frequent despite aggressive therapies<sup>3</sup>. Our patient virtually has all the clinical and histologic features that are in keeping with diagnosis of ECD.

Adult-onset asthma with periocular xanthogranuloma, on the other hand, is frequently associated with lymphadenopathy and elevated levels of polyclonal IgG in addition to its hallmark feature of adult-onset asthma. The distinctive histopathological finding in this disorder is the presence of large lymphoid aggregates with reactive germinal centers<sup>7</sup>.

Adult onset orbital xanthogranuloma presents with isolated orbital lesion without significant systemic involvement such as accompanying immune dysfunction, asthma or paraproteinemia; however, hematological abnormalities such as thrombocytopenia, eosinophilia, anemia and lymphopenia have been anecdotally reported<sup>[4][6]</sup>. As noted by Silva-Call cot et al, there has been a number of reports of orbital xanthogranuloma in adult without significant systemic manifestations termed juvenile xanthogranuloma in an adult or adult onset juvenile xanthogranuloma<sup>[11]</sup>. It is yet not clear whether adult onset xanthogranuloma and juvenile xanthogranuloma in adults are one and same condition or represent different clinical entities. JXG characteristically presents as a single or rarely, multiple yellow and brown skin nodules mostly found on the face, neck and trunk of infants and young children<sup>8,9</sup>. Extracutaneous JXG can affect the eye and ocular adnexa as well as central nervous system, lung, liver, spleen and other sites. Ophthalmic involvement is rather rare and reported to occur in a range of 0.3% to 10% of children with cutaneous JXG<sup>10</sup>. Ocular JXG preferentially affects the iris and can potentially lead to blinding complications such as hyphaema, secondary glaucoma and amblyopia<sup>10</sup>. Orbital involvement is quite uncommon and is often unassociated with skin manifestations as is seen in our cases<sup>6,11</sup>. One of our patients had bilateral flexion deformity of the little fingers of unknown clinical significance.

Our cohort exhibited some characteristics worthy of mention. None of our patient presented with the not uncommon characteristic macroscopic appearance of diffuse, yellow, plaque-like masses in the eyelid that often arouse the clinical suspicion of xanthogranuloma due to the non-superficial nature of the lesions. The possibility of this rare disease should, therefore, be borne in mind even in the absence of characteristic lesions in the eyelid. The differential diagnosis for orbital xanthogranulomatous lesion from clinical point of view is broad and include lymphoproliferative disorder, thyroid eye disease, idiopathic orbital inflammatory disease, Rosai-Dorfman disease etc.<sup>4</sup>. Also, of note is the presence of bony destruction in the two patients with AOX. Previous reports postulated that bony destruction is an unlikely feature of adult onset xanthogranuloma but juvenile xanthogranuloma. One recent report also found bony destruction in patient with ECD<sup>2</sup>. The bony destruction seen in the two cases of AOX in this report may likely be due to continued growth of the xanthogranulomatous lesion over a long period of time as both cases presented years (3 and 5 years) after onset of symptoms. The partly viscous nature of the lesion in these patients with AOX suggest that the lesion may have undergone some form of liquefactive necrosis. The yellowish discoloration of the intra-orbital lesions in AOX in the current series have not been reported before.

Management of orbital xanthogranuloma is currently empirical with variable outcomes. Conservative management is an option if the patient is asymptomatic but issues such as proptosis, diplopia, orbital pain, mechanical ptosis, and cosmetically disfiguring skin lesions necessitate treatment<sup>2</sup>. Various treatment options in use alone or in combination for treatment of these lesions or any underlying hematologic or systemic abnormality

include surgery, intralesional and/or systemic steroids, chemotherapy, radiotherapy, plasmapheresis etc.<sup>5</sup>

Surgical excision is reported by some to be associated with high risk of short term (6 to 12 months) recurrence<sup>1, 12</sup>. All our five patients underwent surgical excision and were followed up for at least one year with maximum being 10 years. Only in one patient with juvenile orbital xanthogranuloma did we have bilateral recurrence 8 years later. Our experience is similar to report by Sivak-Callcott et al of surgery alone being effective in six of their eight patients with AAPOX, although duration of follow up was not mentioned. Davies et al reported 50% recurrence rate following surgery occurring 2 years after in their patient with AOX and 9 years in the one with AAPOX.

Steroids such as oral prednisolone has been shown to induce significant reduction in lesion size and symptoms in some cases<sup>2</sup>. However, it is not uncommon to have recurrence of lesion when it is withdrawn<sup>1, 12</sup>. Intralesional corticosteroid injection (triamcinolone acetonide 40 mg/ml) has also been successfully used in controlling symptoms and signs of AOX and occasionally NBX with eyelid and orbital involvement however they are generally less efficacious than systemic steroids<sup>4, 12</sup>. In cases refractory to systemic corticosteroid therapy, some reports showed methotrexate was efficacious in preventing recurrence of AOX and AAPOX<sup>2, 13</sup>. Systemic steroids, radiation therapy, and chemotherapy, including cyclophosphamide, doxorubicin, and vincristine has been used for treatment of ECD with variable outcomes<sup>4</sup>. Alpha - Interferon and cladribine has also been used with good response and clinical improvement in some reports<sup>14, 15</sup>. Retro-orbital irradiation is often not effective<sup>4</sup>. However, administration of radiotherapy to the orbit in combination with systemic corticosteroids has proven successful in

some cases of orbital xanthogranulomatous diseases. This approach is thought to be more efficacious than surgery, although exacerbation of cutaneous lesions after treatment has been reported<sup>12</sup>. In recent times, BRAF inhibitor (vemurafenib, dabrafenib) is been used in the treatment of ECD with promising results. Its use stems from the identification of BRAF mutation as a possible etiologic factor in ECD<sup>2, 12</sup>. Our patient with ECD was started on oral prednisolone without significant improvement in the marked bilateral exophthalmos, chemosis and conjunctival hyperemia that he presented with. After bilateral surgical debulking of the orbital lesions and continued systemic steroid, his clinical condition improved remarkably. However, his poor vision of light perception from a combination of bilateral compressive optic neuropathy and exposure keratitis persisted.

In conclusion, Orbital xanthogranuloma is a rare heterogeneous group of diseases with similar histopathologic profile. Diagnosis of the disease is made by considering both the clinical and histopathological features. Bony destruction can occur in adult onset xanthogranuloma due to the continued growth of the lesion. Both bony erosion and yellowish discoloration of orbital lesions have not been reported before in AOX. Treatment of orbital xanthogranulomatous disease at present is largely empirical with variable outcomes.

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