



Research Article

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Identification of Stromal Corneal Dystrophy and its Prevalence in the Population of Lahore, Pakistan

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ABSTRACT: *Stromal Corneal Dystrophy (SCD) is a heterogeneous group of inherited corneal disorders that result in corneal opacities, caused due to genetic mutations leading to the accumulation of insoluble material in the cornea. A cross sectional study was conducted by visiting seven main hospitals having higher patients' turnover for eye-check to identify Stromal Corneal Dystrophy and its prevalence among the general population residing in Lahore, main city of Pakistan with total population of 11.13 million according to 2017 census Pakistan. The SCD was determined in patients with respect to age and gender and SCD sub-types. The patients were subjected to a visual acuity test using Snellen chart. Out of 4200 patients, the total cases 107 patients (2.5%) were detected with SCD including 62 (57.94%) cases of males and 45 (42.05%) cases of females and other cases were related to other types of corneal dystrophy which were bilateral and not considered further. The prevalence was higher in the young age group (21-30 years) including 31 patients both male and female. Among the SCD sub-type, Granular Corneal Dystrophy was noticed as a most common type of SCD, affecting 32% of patients. It was concluded both males and females were equally affected with SCD however; its prevalence was high in the young population.*

Key words: *Stromal Corneal Dystrophy, Granular Corneal Dystrophy, Identification, Prevalence, Awareness, Lahore*

INTRODUCTION

Visual acuity refers to the clarity of vision and is important to our daily life. Corneal Dystrophies are group of heterogenous disorders which are characterized by the gradual loss of clarity of the cornea arising from the accumulation of deposits within the various layers of the cornea (Poulaki and Colby, 2009; Weiss et al., 2015). This is mainly an inherited disorder and runs in families. The dystrophy when occur affects eyes bilaterally, progresses steadily with age, and is not affected by environmental or systemic factors (Afshari, 2014). Corneal dystrophies are one of the key diseases in the eastern and western world of corneal clarity (Rahman et al., 2009). According to two sub-classifications, anatomical-histological group distinguishes dystrophies by their presence in the affected corneal layers. They are granular, amyloid, lattice, mosaic, etc. (Bourges, 2017).

Stromal Corneal Dystrophy affects the stromal and central layer of the cornea and spreads into the anterior corneal layers after time, some of which can affect the membrane of descemet and the endothelium. Other layers of the cornea will advance to affect some of these disorders. Stromal Corneal Dystrophy is sub-divided into Macular

Corneal Dystrophy, Granular Corneal Dystrophy, Avellino Corneal Dystrophy, Lattice Corneal Dystrophy, Schynder Corneal Dystrophy, Fleck Corneal Dystrophy and Congenital Stromal Dystrophy. Granular corneal dystrophy and macular corneal dystrophy are the most common forms of stromal corneal dystrophy (Klintworth, 2009). Degenerative and hereditary corneal disease is difficult to discriminate. They are classified based on their phenotype, genotype and evidence obtained for their diagnosis by the IC3D classification. Medical procedures consist of drops, ointments, hyperosmotic agents and bandage contact lenses to facilitate healing. Less invasive surgical procedures such as phototherapeutic keratectomy and lamellar keratectomy are used as second-line therapy. The present study was conducted to identify the patients of various forms of stromal corneal dystrophy and its prevalence in different patients and age groups.

MATERIALS AND METHOD

Patients affected with stromal corneal dystrophies were examined by visiting General Hospital, Mughal Eye Hospital, Services Hospital, Mayo Hospital, Shaikh Zayed Hospital, Al Ehsan Welfare Eye Hospital and Layton Rahmatullah Benovolent Trust Hospital (Table 1).

Table 1: Number of Patients in each Hospital of Lahore

Name of Hospitals	Number
General Hospital	11
Mughal Eye Hospital	17
Services Hospital	15
Mayo Hospital	17
Shaikh Zayed Hospital	9
Al Ehsan Welfare Eye Hospital	18
Layton Rahmatullah Benovolent Trust Hospital	21

In order to obtain primary information about the postal address, diagnosis and disease status of patients, perform as were filled out by asking the cornea patients in the hospitals. Hospital records of the corneal patients were assessed. The staff of these hospitals was supportive and corporative and assisted in providing all the data with complete information about the patients of stromal corneal dystrophies.

Variables assessed

Case files of patients with stromal corneal dystrophies were assessed. The variables considered for analysis of patients, were age, gender, Far Visual Acuity, Main complaint, and any associated pathologies.

Clinical evaluations

The diagnosis of stromal corneal dystrophy was based on Visual acuity

test. Visual acuity or clearness of vision is important to our daily life, the most common visual function measurement, visual acuity, is a relatively crude and narrow one that measures only a small part of the large spectrum of visual functions. A common cause of poor visual acuity is refractive error, or defects in how the light is refracted are most widely used to assess visual acuity in the eyeball chart of Snellen. Visual acuity is the calculation of the ability to distinguish toward two objects at high contrast relative to the background, separated in space. This is clinically assessed by asking the subject to discriminate against letters from a specified visual angle as described by Sue (2007). The patients were asked to sit 20 feet (6 meters) away from the Snellen' schart (Fig. 1) and tried to read the alphabets on the chart.

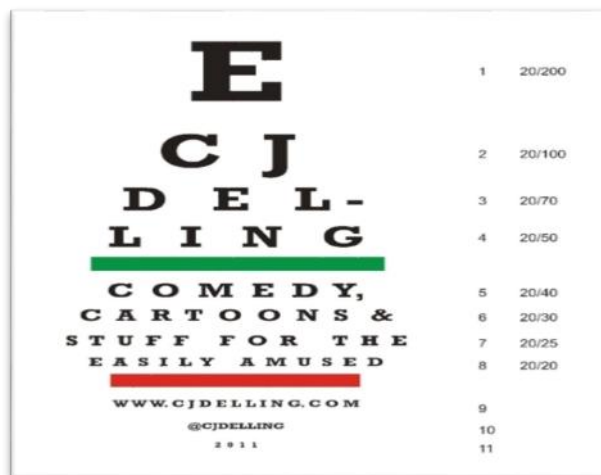


Fig. 1: Snellen Chart (<http://www.cjdelling.com/>)

RESULTS

Among 4200 patients studied from different hospitals of the Lahore, 107 (2.55%) cases of stromal corneal dystrophies were recognized which include 45 cases (42.05%) of females and 62 cases (57.94%) of males. Two hundred and fourteen eyes (90 of female and 124 of males) of 107 patients were assessed and other 4093 patients were related to other types of corneal dystrophy, which were posterior, anterior and

epithelial corneal dystrophy so they ignored. Stromal Corneal Dystrophies were found to be bilateral in 190 eyes and isolated in 24 eyes. The most common age group found was 21- 30 years with 31patients. While, 20 patients were categorized in the age group of 31- 40 years. There were 17 patients in age group of 11- 20 years, 20 patients with age group of 41- 50 years, 9 patients with age group of 51- 60 years, 6 patients with age group of 61- 70 years and 4 patients with age group of 0- 10 years (Table 2).

Table 2: Number of Patients in different Age Groups

Age Groups in Years	No. Of Patients
61- 70	6
51- 60	9
41- 50	20
31- 40	20
21- 30	31
11- 20	17
0- 10	4

Main complaint

Main complaint by all patients was that their visual acuity was lost gradually. From total 107 Patients of SCDs, pain was observed in the eyes of 83 (70%) patients in which 48 (44.85%) patients were males and 35 (32.7%) patients were females. Refractive error was examined in 214 eyes from which 45 were female eyes and 62 were male eyes. The most common refractive error was a very common eye disease. It occurs when the eye cannot clearly see the images from the outside world. The result of refractive errors is blurred vision, which is sometimes causes visual impairment, found was 6/24.

examined and Snellen readings were recorded. Only 8.41% patients (n = 9) were found to have normal visual acuity (6/6). Moderate vision loss (6/12, 6/15, 6/18, 6/20, 6/21, 6/22, 6/23, 6/24 and 6/25) was recorded in 37.4% (n = 40) patients while high vision loss (6/29, 6/30, 6/32, 6/35, 6/36, 6/39, 6/40, 6/42, 6/43, 6/50, 6/55 and 6/60) was recorded in 43% (n = 68) patients, respectively (Fig. 2).

23 patients out of 107 patients were of Macular Corneal Dystrophy (MCD) among which only 21.7% (n = 5) patients have normal vision. There were 30.4% (n = 7) patients with moderate vision loss and 47.8% (n = 11) patients with high vision loss.

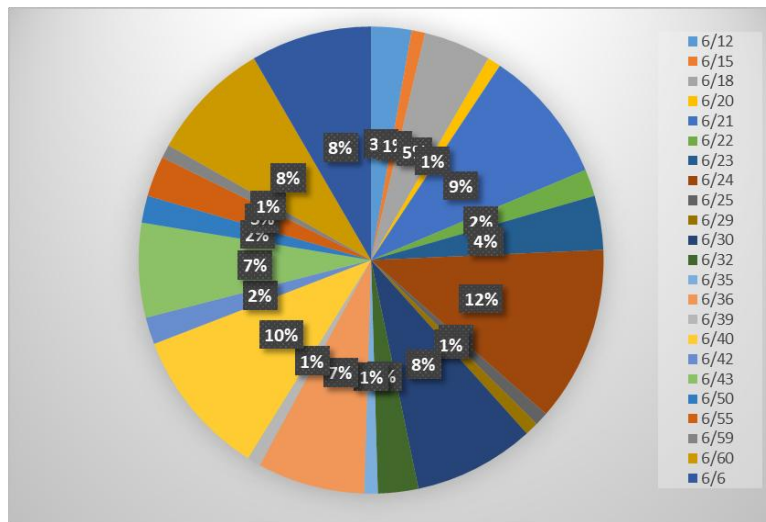


Fig. 2: Prevalence of different types of stromal corneal dystrophy by age groups

Prevalence of different types of Stromal corneal dystrophy according to visual acuity

214 eyes of 107 patients suffering from Stromal Corneal Dystrophy were

patients with high vision loss. 35 patients out of 107 were of Granular Corneal Dystrophy (GCD) among which only 8.57% (n = 3) patients had normal vision, 40% (n = 14) patients have moderate vision

loss and 51.4% (n = 18) patients had high vision loss. 26 cases were found to be of Lattice Corneal Dystrophy (LCD) among which 23.07% (n = 6) patients had moderate vision loss, 73.07% (n = 19) patients had high vision loss and one patient have normal visual acuity. Out of 107 patients, 6 patients were of Avellino corneal Dystrophy (ACD) among which 50% (n = 3) patients had moderate vision loss and 50% (n = 3) patients had high vision loss. 10 patients were of Congenital Stromal Corneal Dystrophy (CSCD), among which 70% (n = 7) patients had moderate vision loss, 2 patients (20%) had high visual loss and 1 (10%) patient had normal visual acuity. 7 out of 107

patients were of schnyder corneal dystrophy among which 5 patients (71.43%) had higher vision loss and 2 patients (28.6%) had less vision loss. Our study reported some major dystrophies and their percentages were Macular (21.5%), Granular (32.7%), Schnyder (6.54%), Lattice (24.3%), Avellino (5.6%) and Congenital (9.3%) out of 107 SCD cases.

Prevalence of Stromal Corneal Dystrophy in both gender by age

Out of 107 patients, observed 42.05% (45 cases) were females and 57.94 % (62 cases) were males (Fig. 3).

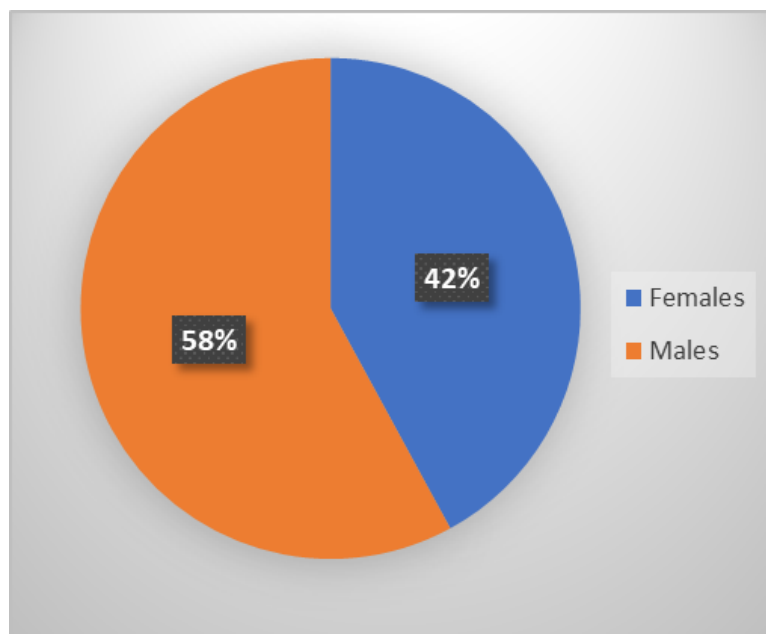


Fig. 3: Prevalence of different types of stromal corneal dystrophy by gender

There were 8 cases of males and 1 case of female found to be affected in the age group of 51-60 years. There were 3.74% patients (1 female and 3 males) with age range of 0-10 years, 15.89% (7 females and 10 males) patients with age range in 11-20 years was found, 28.9% (12 females and 19 males) patients fall in the age group of 21-30 years, 18.7% (9 females and 11 males) patients fall in the age group of 31- 40 years, 18.7% (11 females and 9 male) patients present in the age group of 41- 50 years. Only 5.6% (2 males or 4 females) patients were found to be affected in the age group of 61 -70. Out of 107 cases of SCD, 35 (33.33%) patients were analyzed as GCD from which 45.7% (n = 16) were females and 54.3% (n =19) were of males. In age group of 11- 20 years 11.42% (1 female and 3 males) patients were present. In the age group of 21- 30 years 40% (4 females and 10 males) patients were observed. In the age group of 31-40 years, 20% (4 females and 3 males) patients were examined. 14.3% (3 females and 2 males) patients were present in age group of 41-50 years. In age group of 61-70 years, 8.6% (2 females' and1 male) patients were examined. Out of 107 cases of SCD, 21.5% (n = 23) patients were of MCD from which 4 (17.4%) were females and 19 (82.6%) were males. Only one male (4.35%) fall in age group of 0-10 year. In age group of 11- 20 years, 1 female (4.35%) and 2 males (8.69%) were present. In the age group of 21- 30 years, 2 females (8.69%) and 4 males (17.4%) were observed. In the age group of 31-40

years 4 males (17.4%) were examined. 1 female (4.35%) and 5 males (21.7%) fell in age group of 41-50 years. 3 males (13.04%) fell in the age group of 51-60 years. Out of 107 cases of SCD only 6 cases (5.61%) of Avellino Stromal Corneal Dystrophy were analyzed from which 2 (33.33%) were females and 4 (66.66%) were males. In age group of 11-20 years only male (16.6%) is present. 1 female (16.6) was present in age group of 41-50 years. In age group of 51- 60 only 3 males (50%) were present. In age group of 61-70 years only1 female (16.6%) was examined.

DISCUSSION

Corneal dystrophies are considered to affect corneal clarity and refraction as dominant, hereditary, bilateral disorders, resulting in varying degrees of visual disruption. They are usually said to be early, axial, symmetrical, steadily progressive, vascularization-free and not correlated with other systemic conditions (Hemadevi et al., 2017). In our study, we observed 107 cases of Stromal Corneal Dystrophy from which 42.05% were females and 57.94% were males.24 out of 107 were isolated i.e. they occur in single eye and remaining 190 were bilateral. Mostly corneal dystrophies were bilateral as reported by Poulaki et al. (2008). Dystrophies are categorized into Fuchs dystrophy, Lattice dystrophy, Granular dystrophy, Macular dystrophy and others (20%) according to the French National Waiting List (Poinard et al., 2009). Our

study reported some major dystrophies and their percentages were Macular (21.5%), Granular (32.7%), Schnyder (6.54%), Lattice (24.3%), Avellino (5.6%) and Congenital (9.3%) out of 107 SCD cases. This study will help in updating the predominance of stromal corneal dystrophy in Pakistan. Proper clinical assessment, early finding, genotyping, genetic counseling and proper management are needed for the restoration of vision loss due to SCD.

Granular Corneal Dystrophy was key focus on the occurrence of Stromal Corneal Dystrophies and their manifestations, the most abundant dystrophy found. In the corneal stroma, GCD is distinguished by distinct opacities that are irregular or flak like, appearing somewhat whitish or glassy. While most patients are asymptomatic, persistent erosion occurs in some patients. Over time, the lesions become more numerous and sever, leading to reduction in visual acuity. Some patients in the fifth decade or later can need keratoplasty (Joel et al., 2009).

Predominance of Macular Corneal Dystrophy is not known. Instances of MCD have been distinguished around the world. This state is normal in India, Iceland, Saudia and in some regions of the USA (Klintworth, 2006). Despite the fact that MCD is less normal throughout the world than LCD or GCD, it is the most well-known of the corneal stromal dystrophies in spots, for example, Iceland and Saudi Arabia

(Weiss et al., 2015). There were 100 eyes of MCD patients examined and visual acuity of all the eyes (100%) was recorded. It was observed that most of the patients have blur vision and high vision loss. Moderate vision loss (6/21, 6/23 and 6/24) was recorded in only 13 eyes (13%) out of 100. Rest of the eyes had high vision loss (6/30, 6/36, 6/40, 6/42, 6/43 and 6/60) thus was observed in 87 eyes (87%). No patient was found to have normal visual acuity (Naz et al., 2018). According to our study the Macular Corneal dystrophy most common dystrophy after the Granular Corneal dystrophy and Lattice Corneal dystrophy. The most representative age group in Macular Corneal Dystrophy was 21- 40 years. Out of 23 patients of Macular Corneal Dystrophy only 21.7% (n = 5) patients have normal vision.

Lattice corneal dystrophy was known to be the most common form of stromal corneal dystrophy. Autosomal dominant, bilateral disorder usually presents with symptoms of persistent corneal erosion a reduced vision during teenage. Some types of LCD are present in early life (Kannabiran et al., 2006). The prevalence of LCD is 0.32% out of 2213 patients of corneal disease (Bhatti et al., 2011). According to our study the Lattice Corneal Dystrophy was second most common type. The prevalence of LCD was 24.29% out of 107 patients of SCD. Out of 107 cases of SCD 26 cases of Lattice SCD were analyzed from which 15 were females and 11 were males. In 21-30 years of age group, 3 females

(16.66%) and 6 males (33.33%) were observed. 6 females (33.33%) and 3 males observed in age group of 31-40 years.

Avellino is also type of Granular Corneal Dystrophy but it is less common. According to the Lee et al., (2008), one out of 870 individuals is at risk of carrying Avellino Corneal Dystrophy gene. In our study, the prevalence of Avellino was 11% out of 107 individuals of SCD. We identified 6 patients of Avellino from which 50% patients lie in group of 51-60 years of age, 17% lie in group of 61-70 years of age, 17% in age group of 41-50 and 17% lie in age group of 11-20 years.

Congenital stromal corneal dystrophy (CSCD) is less prevalent and is distinguished by the various opaque flaky or feathery clouding of the corneal stroma than the other dystrophies (Van et al., 2002). In some of the large French and Norwegian families, affected individuals with CSCD have been extensively studied. The prevalence of CSCD in our studies was 9.34% (n=10) out of 107 patients SCD patients, and it was less frequent than LCD, MCD and GCD. The bulk of the representative age group was 11-20.

According to Lee et al., (2010), one out of 870 individuals was at risk of carrying that Schnyder Corneal Dystrophy (SCD). SCD as a global condition found in the major ethnic and racial groups. Although prevalence is supposed to be same, the actual rates from country to country will be better understood once representative test numbers are completed

(Lee et al., 2010). Schnyder Corneal Dystrophy is less common disease. In our study, the prevalence of Schnyder dystrophy was 6.54%. Out of 7 cases of Schnyder 28.57% lies in age group of 40-50 years.

CONCLUSION

In this study, 107 cases of Stromal Corneal Dystrophy were observed and prevalence was high in males as compared to females. Our study reported some major dystrophies, and their percentages were macular (21.5%), granular (32.7%), schnyder (6.54%), lattice (24.3%), avellino (5.6%) and Congenital (9.3%) out of 107 SCD cases. This study will help in updating the predominance of stromal corneal dystrophy in Pakistan and particularly be valuable in providing awareness in affected individuals, which will help in reducing total burden of disease. Proper clinical assessment, early finding, genotyping, genetic counseling and proper management are needed for the restoration of vision loss due to SCD.

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