Chapter V

Results

RESULTS

The population of the study included 30 unrelated patients with retinitis pigmentosa (RP) (probands). All cases were studied prospectively (10/1985 — 11/1988). To identify the genetic type of the RP patients, their relatives including parents; grandparents, siblings and offsprings were examined whenever possible.

For evaluating the value of fluorescein angiography in identifying the disease in patients with no eye ground changes, though at high risk for the development of RP, an additional case was seen in "AEI".

GENETIC ANALYSIS: (Table X, Fig. 4)

Among the 30 RP patients, the family history of 7 was negative and indicated no specific hereditary mode, those were termed simplex (S). They constituted 23.4% of the study population, the second largest group.

Those with a definable hereditary transmission included 19 patients (63.3%) autosomal recessive (AR), 2 patients (6.7%) autosomal dominant (AD), and one patient (3.3%) sex linked (XL). Only one case fitted the undetermined genetic category (UG). The latter case was a proband in

Table (X): Distribution of RP Patients by Genetic Type.

Genetic type	Patients	
	No.	%
Autosomal dominant (AD)	2	6.7
Autosomal recessive (AR)	19	63.3
X-linked recessive (XL)	1	3.3
Simplex (S)	7	23.4
Undetermined genetic (UG)	1	3.3
Total	30	100.0

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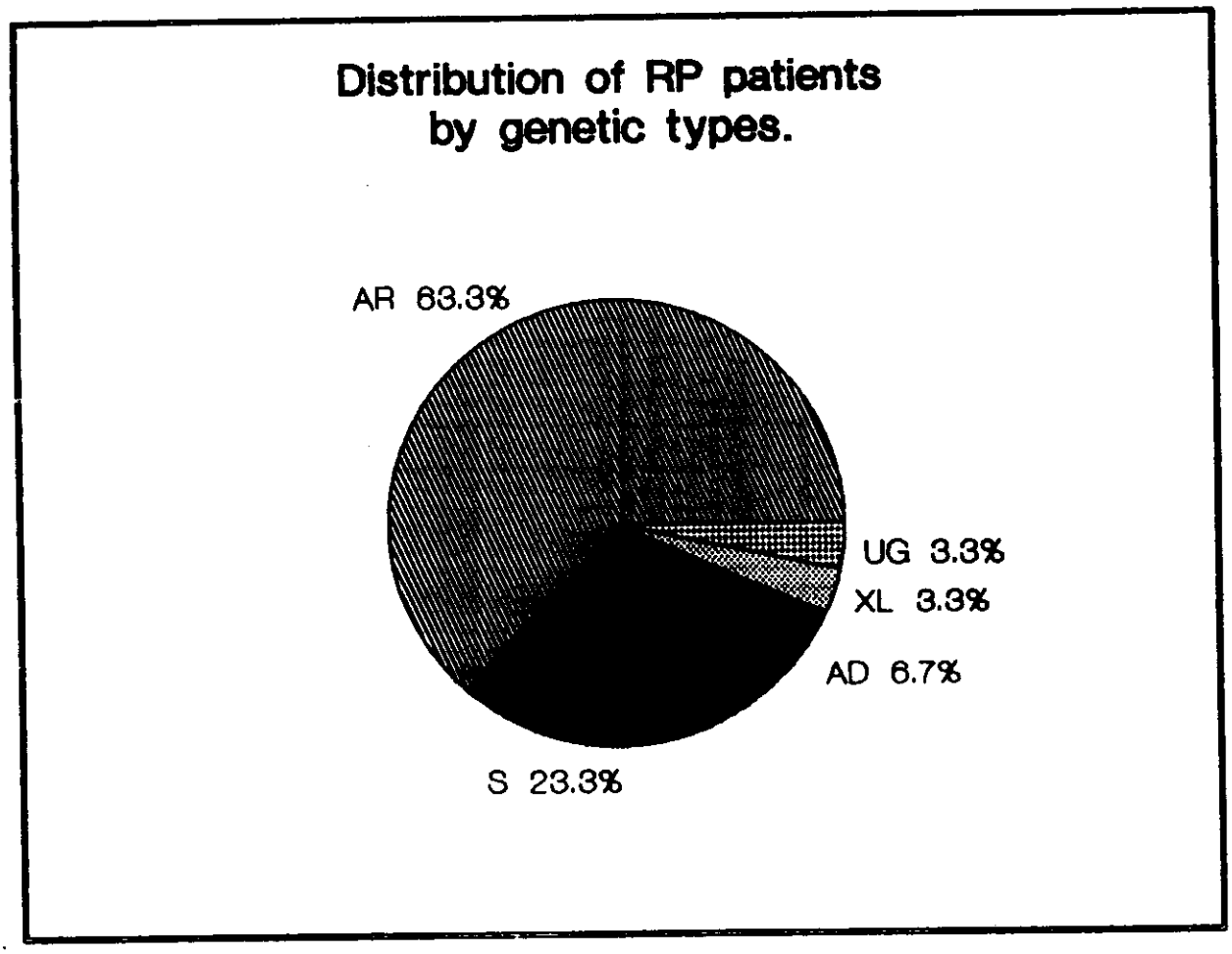

Fig.(4): Distribution of RP patients by genetic types.

a male multiplex group with negative parental consanguinity. This may be AR or XL.

Among the 19 AR patients, 2 were diagnosed as Usher's syndrome (RP with congenital deafness).

N.B.: Examples of pedigrees of RP patients with different genetic types of inheritance are shown in figures 5-8.

SEX AND AGE DISTRIBUTION IN RP PATIENTS:

Among the 30 RP patients (probands), 20 patients were males and 10 patients were females. The male to female ratio was 2:1 with a male incidence of 66.7% (Table XI, Fig.9). male incidence was higher than female among all genetic groups (Table XII).

The age of RP patients ranged from 18-53 years with a mean age of 34.56±9.11 years (Table XII). There was no statistically significant difference between the mean ages in the different genetic types, where $F = 1.9599$. Out of the 30 patients constituting this series, 56.7% were more than 32 years old.

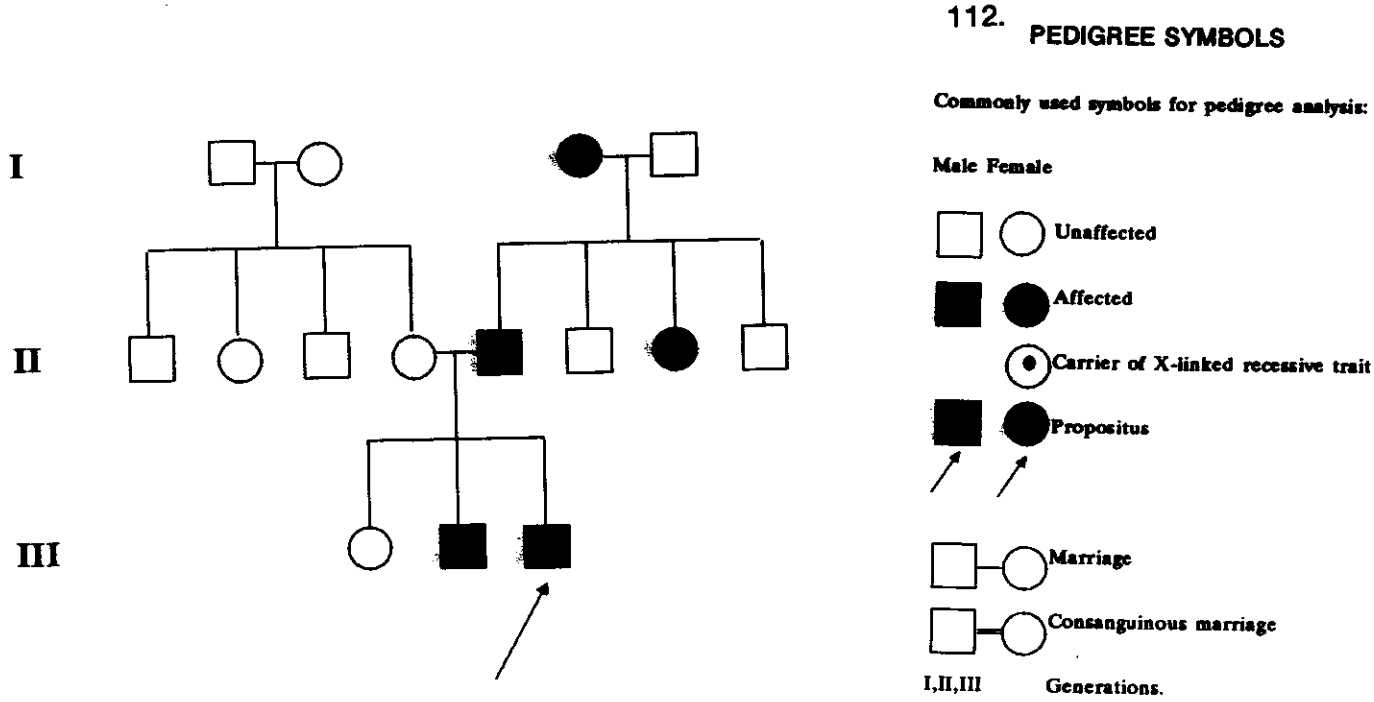

Fig.(5): A pedigree of an autosomal dominant (AD) family.

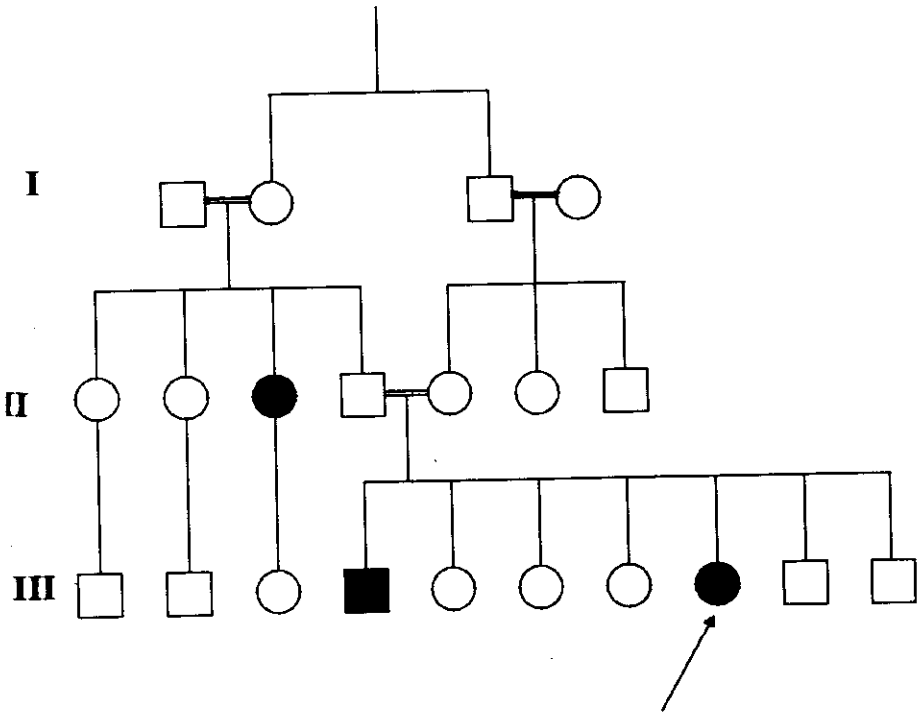

Fig.(6): A pedigree of an autosomal recessive (AR) family.

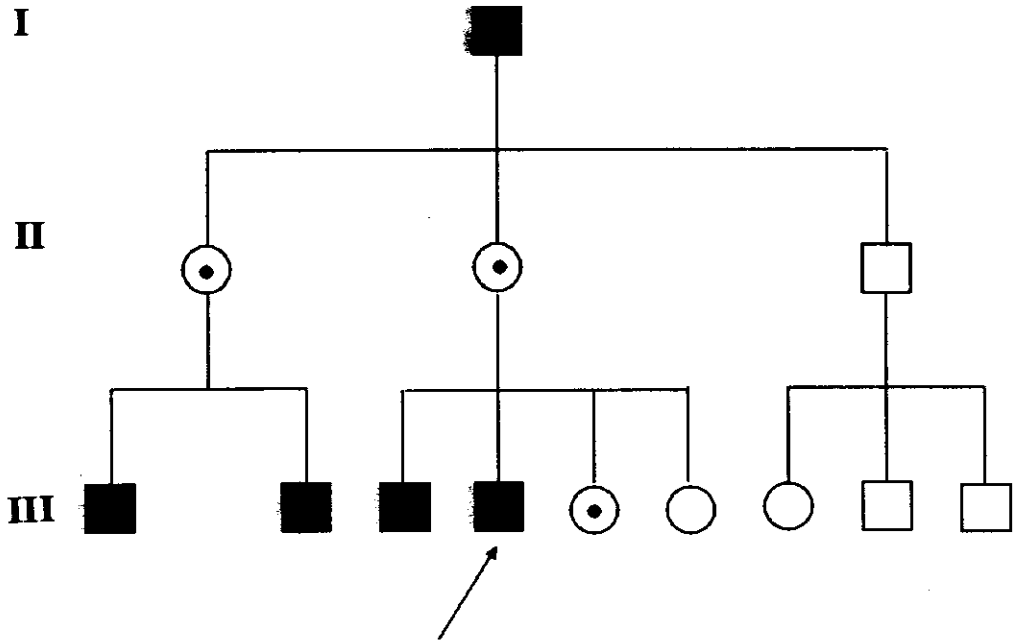

Fig.(7): A pedigree of a sex-linked (XL) family.

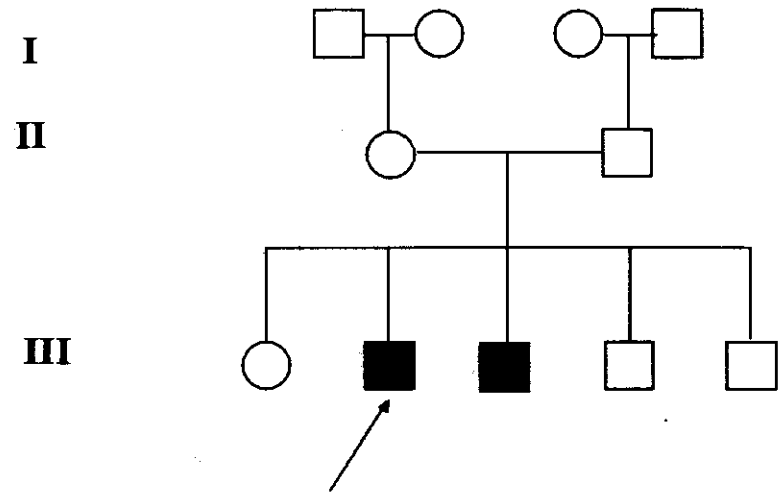

Fig.(8): A pedigree of an undetermined genetic (UG) family.

Table (XI): Sex Distribution in Cases of RP (Probands).

Genetic type	Patients	
	No.	%
Male	20	66.7
Female	10	33.3
Total	30	100.0

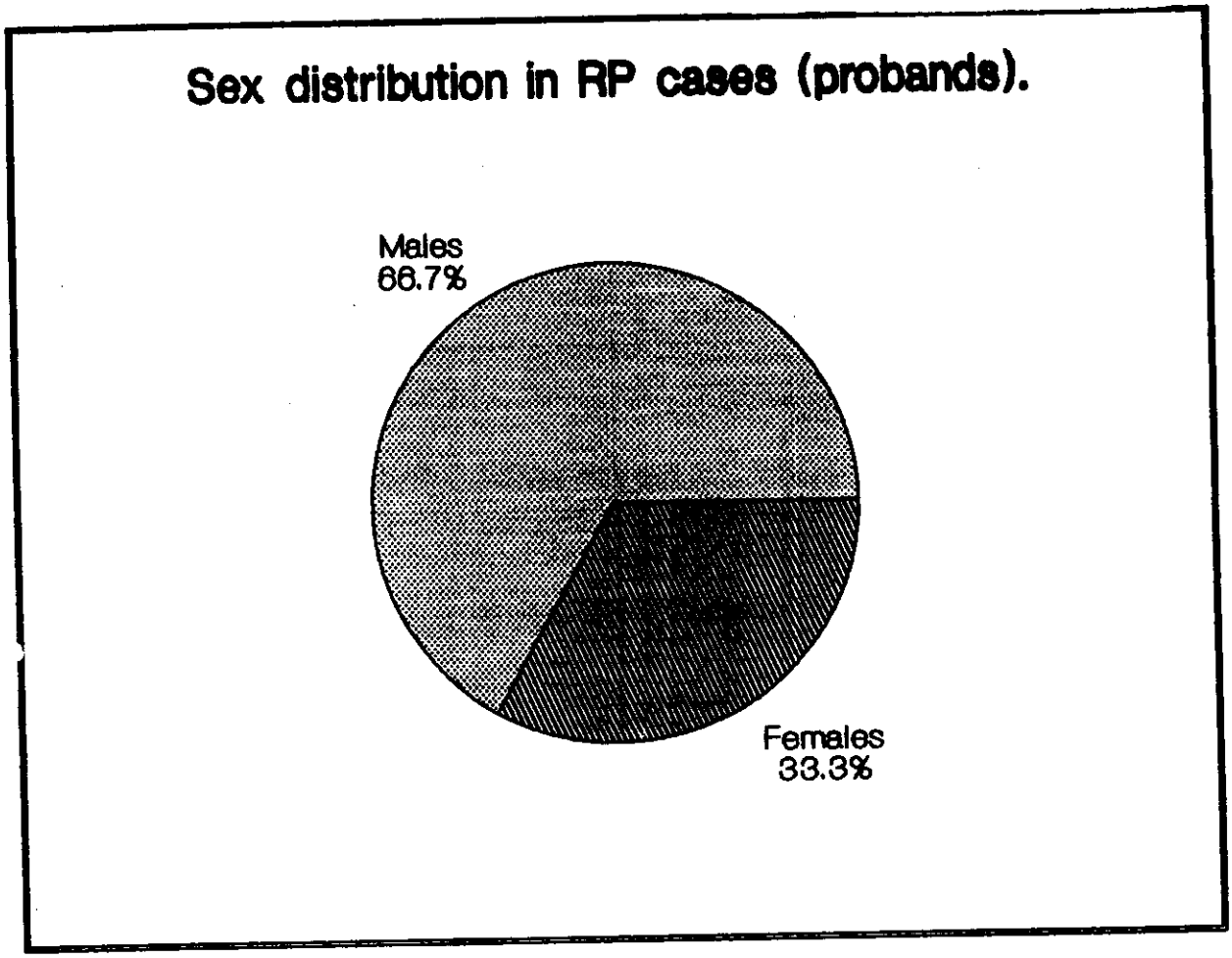

Fig.(9): Sex distribution in RP cases (Probands).

Table (XII): Sex and Age Distribution in the Different Genetic Types.

	Genetic type					
	AR	S	AD	XL	UG	Total
	19 (63.3)	7 (23.4)	2 (6.7)	1 (3.3)	1 (3.3)	30
Sex						
Male	12 (63.2)	4 (57.1)	2 (100)	1 (100)	1 (100)	20
Female	7 (36.8)	3 (42.9)	-	-	-	10
Age (Ys)						
Range	18-52	25-50	29-36	33	53	18-53
$\overline{X}$	32.32	38.85	32.5	33	53	34.56
"S"	8.62	8.87	4.95	0	0	9.11

* Numbers indicate number of patients, and numbers in parenthesis are percentages.
* "S"= Standard Deviation.

PARENTAL CONSANGUINITY IN RP PATIENTS:

Parental consanguinity was observed in 66.7% of RP patients (probands), with 56.7% of the cases of consanguinity being of the first cousin order (Table XIII). Parental consanguinity was present in 100% of the AR patients.

SYMPTOMS:

All cases were complaining of night blindness and it was the first complaint in 100% of cases. Other relevant symptoms were visual field affection, diminution of vision and prolonged dark adaptation.

As regards the age of onset of symptoms (Table XIV), in 56.7% of the RP patients, the onset was in the first decade of life, while 96.7% of patients became symptomatic by the age of 20 years. The corresponding figures in the AR group were 63.2% and 94.8% respectively. There was no significant difference between the mean decade of onset of all studied genetic types where $F = 0.4373$.

VISUAL ACUITY IN RP PATIENTS IN RELATION TO MODE OF INHERITANCE:

The percentage of eyes of RP patients with good vision (6/18 or better) constituted 28.3% of the studied population, whereas the

Table (XIII): Parental Consanguinity in RP Patients.

Parental consanguinity	Patients	
	No.	%
1st cousins	17	56.7
Other positive	3	10
Negative	10	33.3
Total	30	100.0

Table (XIV): Age of Onset of Symptoms in Different Genetic RP Types.

Genetic type	Age of Onset (Decade)			Total	$\bar{X}$	"S"
	1st	2nd	– 3rd			
AD	1 (50%)	1 (50%)	-	2 (100%)	1.5	0.71
AR	12 (63.2%)	6 (31.6%)	1 (5.2%)	19 (100%)	1.42	0.61
XL	-	1 (100%)	-	1 (100%)	2	0
S	4 (57.1%)	3 (42.9%)	-	7 (100%)	1.43	0.53
UG	-	1 (100%)	1 (3.3%)	30 (100%)	1.47	0.57

percentage of eyes with affected vision (< 6/18) (low and blind) constituted 71.7% (Table XV).

There was no statistically significant difference between the AR and S groups (2 largest groups) as regards corrected visual acuity in the 3 categories; good (Z = 1.3216), low (Z = 0.6852) and blind (Z = 0.4005).

VISUAL ACUITY IN RP PATIENTS IN RELATION TO AGE GROUPS: (Table XVI)

RP patients more than 32 years old constituted 56.7% of the studied population. Seventy per cent of the above mentioned age group had affected vision (< 6/18) (low and blind). On the other hand, 70% of patients with good vision (6/18 or more) were aged 32 years or less (Fig. 10). This was found to be statistically significant where Fisher's Exact test P = 0.0451.

ERRORS OF REFRACTION IN RP PATIENTS: (Table XVII)

Among the 60 eyes tested, myopia was the most prevalent ametropic condition. It was present in 60% of the eyes of RP population and 63.2% of eyes of AR patients. Of the myopic eyes, 66.7% were less than -6 dioptre.

Table (XV): Distribution of Corrected Visual Acuity Among a Series of 60 Eyes Having RP, by Genetic Type.

Visual acuity	Genetic type					
	AD	AR	XL	S	UG	Total
Good vision						
6/18 or better	1 (25%)	14 (36.8)	-	2 (14.2)	-	17 (28.3)
Low vision*						
<6/18-6/60	3 (75%)	11 (28.9%)	2 (100%)	2 (14.2%)	-	18 (30%)
<6/60-3/60	-	2 (5.3%)	-	6 (42.9%)	-	8 (13.3%)
Blindness						
<3/60-1/60	-	1 (2.6%)	-	-	-	1 (1.6%)
<1/60	-	10 (26.3%)	-	4 (28.6%)	2 (100%)	16 (26.7%)
No. PL	-	-	-	-	-	-
Total	4 (100%)	38 (100%)	2 (100%)	14 (100%)	2 (100%)	60 (100%)

Table (XVI): Visual Affection in Relation to Age Group Among A Series of 30 RP Patients.

Corrected visual acuity	Age group		Total
	⩽ 32 years	> 32 years	
Good			
6/18 or better	7 (53.8%)	3 (17.6%)	10 (33.3%)
Affected			
<6/18-6/60	2 (15.4%)	6 (35.3%)	8 (26.7%)
<6/60	4 (30.8%)	8 (47.1%)	12 (40%)
Total	13 (100%)	17 (100%)	30 (100%)

> = More than.

⩽ = Less than or equal to.

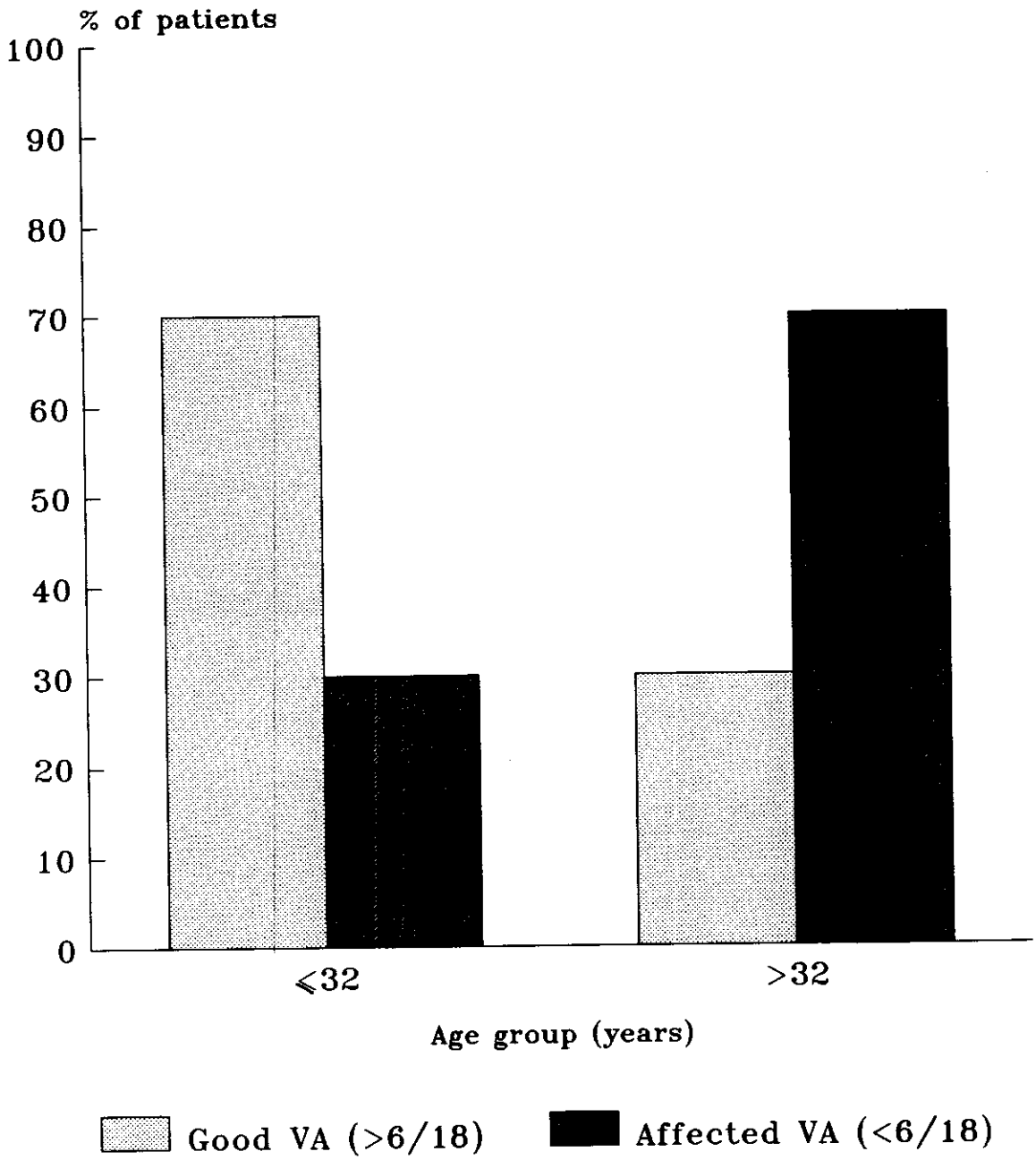

Fig.(10): Distribution of visual affection according to age group.

Table (XVII): Distribution of Refractive Errors (Using Spherical Equivalent) Among a Series of 60 Eyes Having RP, by Genetic Type.

Refractive Errors	Genetic type					
	AD	AR	XL	S	UG	Total
Myopia	2 (50%)	24 (63.2%)	2 (100%)	6 (42.9%)	2 (100%)	36 (60%)
Emmetropia	-	2 (5.3%)	-	3 (21.4%)	-	5 (8.3%)
Hypermetropia	2 (50%)	12 (31.5%)	-	5 (35.7%)	-	19 (31.7%)
Total	4 (100%)	38 (100%)	2 (100%)	14 (100%)	2 (100%)	60 (100%)

The spherical refractive error ranged from +4.25 to -12 dioptre (using spherical equivalent).

There was no significant difference between the AR and S groups as regards the incidence of myopia ($Z = 1.3144$), emmetropia ($Z = 1.7453$) and hypermetropia ($Z = 0.2865$).

VISUAL FIELD CHANGES IN RP PATIENTS:

In this study, visual field was plotted for only 22 out of the 30 RP patients. The field was not plotted in 8 patients due to poor vision ($< 1/2/60$). Also, it was plotted though irreliable in an RP patient with Usher's syndrome. Therefore, the actual number of patients with plotted and reliable fields was 21.

Almost all plotted fields showed generalized contraction to variable extents without remaining peripheral islands.

The most common type of field was the tubular together with the severely generalized contracted type, being present in 76.2% of the plotted group.

The visual fields of the same patient were almost equally affected denoting the bilaterality and symmetrical involvement of both eyes.

The method used in scoring the visual field in this study was previously mentioned (material and methods). Only 5 patients (23.8%) out of the 21 RP patients with plotted field fitted group I. The score of the mean visual field in this study was 28.19±17.7 degrees. There was no statistically significant difference between the mean visual field in the different genetic types where F=2.6538 (Table XVIII).

The degree of visual field changes in the different age groups of RP patients is shown in Table XIX. It was found that 80% of patients belonging to group I visual field were aged 32 years or less (<32 years). On the other hand, 87.5% of patients belonging to group II visual field were more than 32 years old (>32 years) (Fig. 11,12). The score of the mean visual field of patients above 32 years=21.33±13.54 degrees, whereas in patients (≤32 years), the same mean=37.33±19.15 degrees. This difference was found to be statistically significant where r=-0.53, $t_{(n-2)}=2.7245$.

N.B.: Examples of the different visual field groups (I,II,III) are shown in figures 13-15.

LENS CHANGES IN RP:

Among the 60 eyes of 30 patients, were 20 eyes (33.3%) with clear lens (Table XX), a cataract was present in the remaining 40 eyes (66.7%) with no genetic group spared.

Table (XVIII): Distribution of Visual Field Changes Among a Series of 21 Patients Having RP, by Genetic Type.

Field changes	Genetic type					
	AD	AR	XL	S	UG	Total
Group I (>40°)	-	5 (38.6%)	-	-	-	5 (23.8%)
Group II (20°-40°)	2 (100%)	5 (38.6%)	-	1 (20%)	-	8 (38.1%)
Group III (<20°)	-	3 (23.1%)	1 (100%)	4 (80%)	-	8 (38.1%)
Total	2 (100%)	13 (100%)	1 (100%)	5 (100%)	-	21 (100%)
$\bar{X}$	32.50	34.62	10	13.4	-	28.19
"S"	3.54	18.27	0	7.92	-	17.70

$\bar{X}$ = mean.
"S"= Standard deviation.

Table (XIX): Distribution of Visual Field Changes Among A Series of 21 Patients Having RP by Age Group.

Field changes	Age group		Total
	⩽ 32 years	> 32 years	
Group I (>40°)	4 (44.4%)	1 (8.3%)	5 (23.8%)
Group II (20°-40°)	4 (44.4%)	4 (33.3%)	8 (38.1%)
Group III (<20°)	1 (11.2%)	7 (58.3%)	8 (38.1%)
Total	9 (100%)	12 (100%)	21 (100%)
X	37.33	21.33	
"S"	19.15	13.54	

> = More than.

⩽ = Less than or equal to.

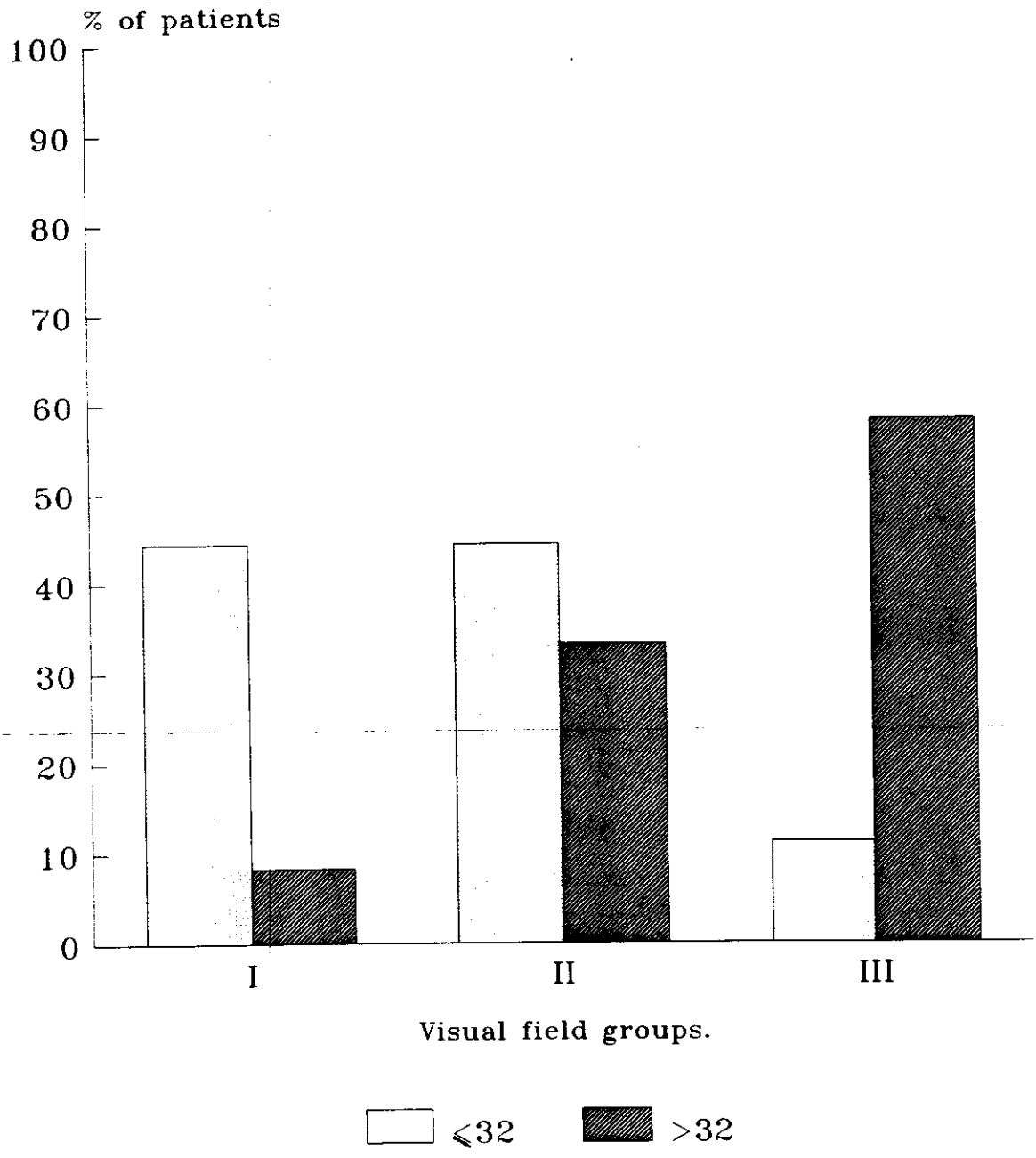

Fig.(11): Distribution of visual field changes according to age group.

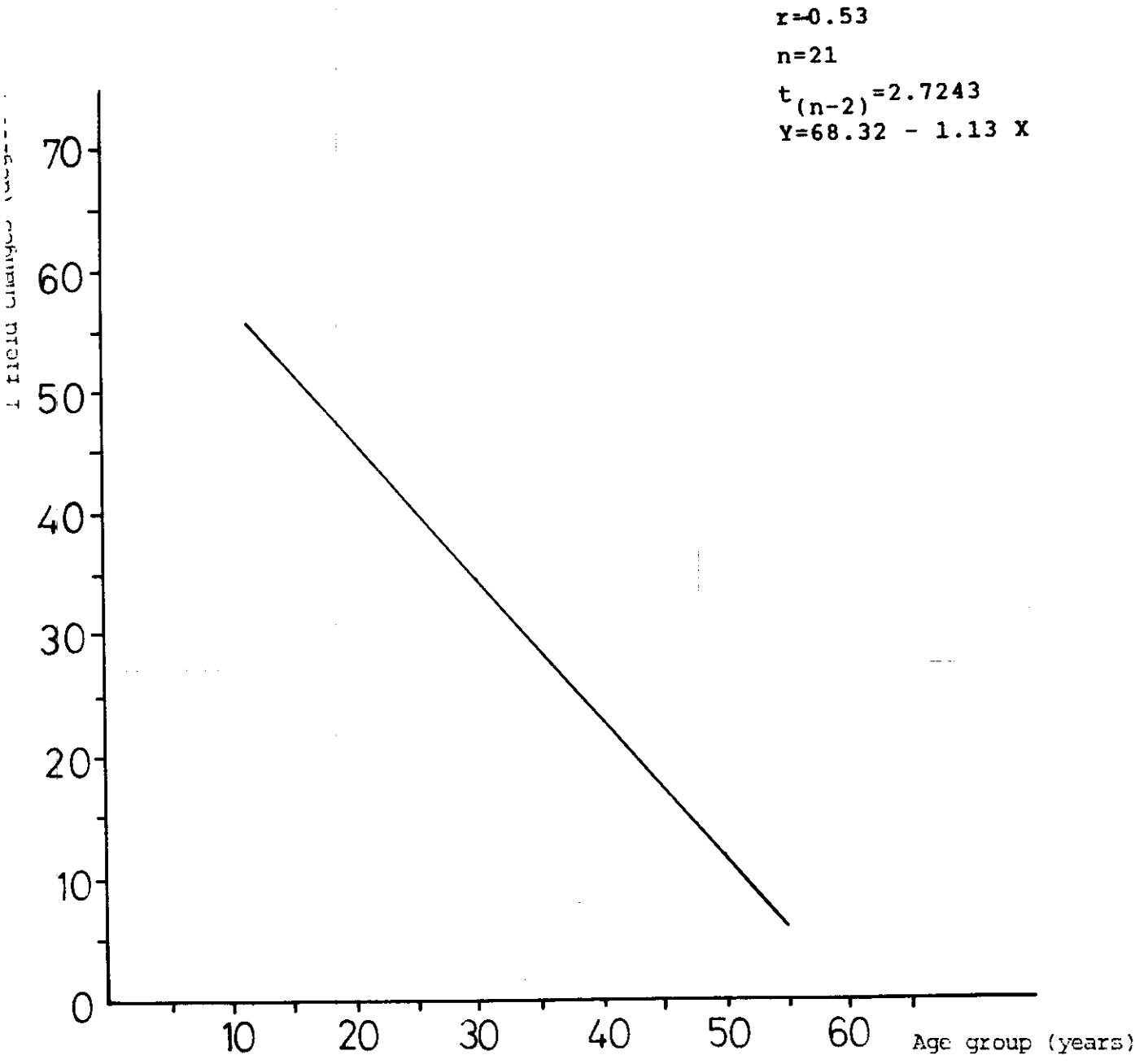

Fig.(12): Correlation between visual field changes (degrees) with age (years).

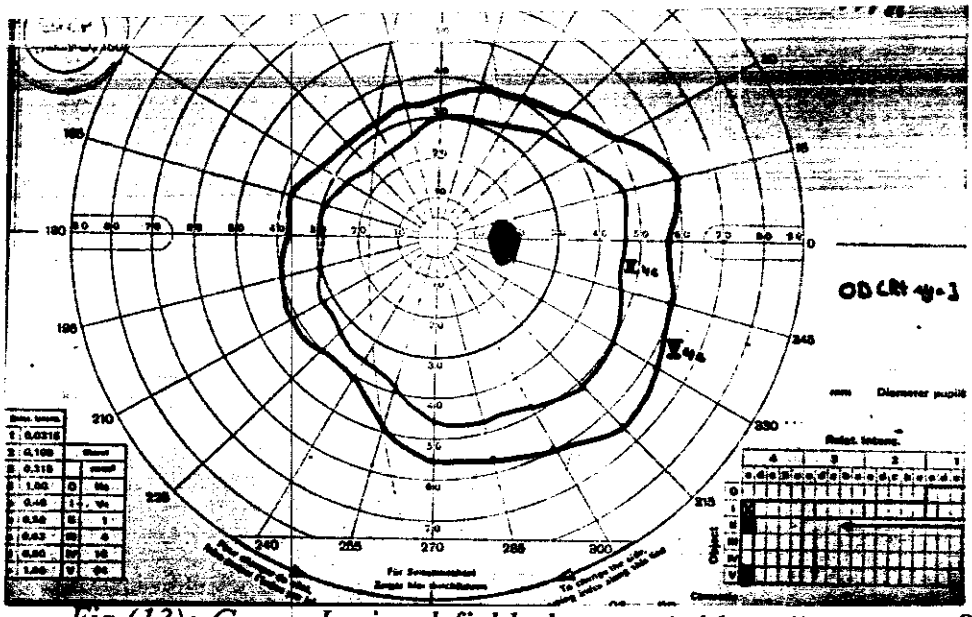

Fig.(13): Group I visual field changes (widest diameter >40°).

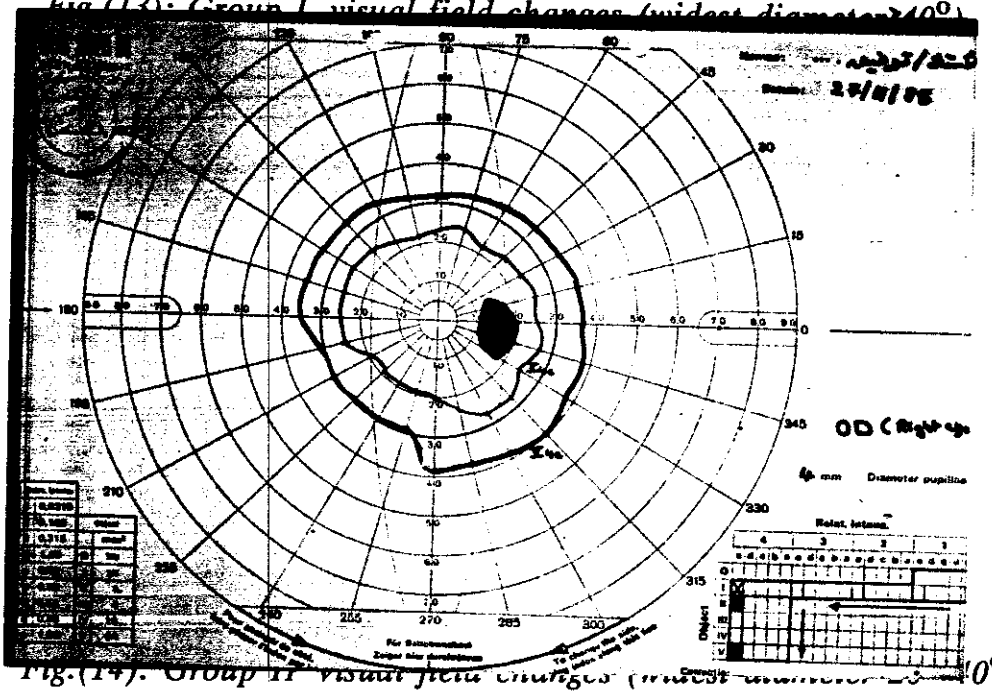

Fig.(14): Group II visual field changes (widest diameter 20-40°).

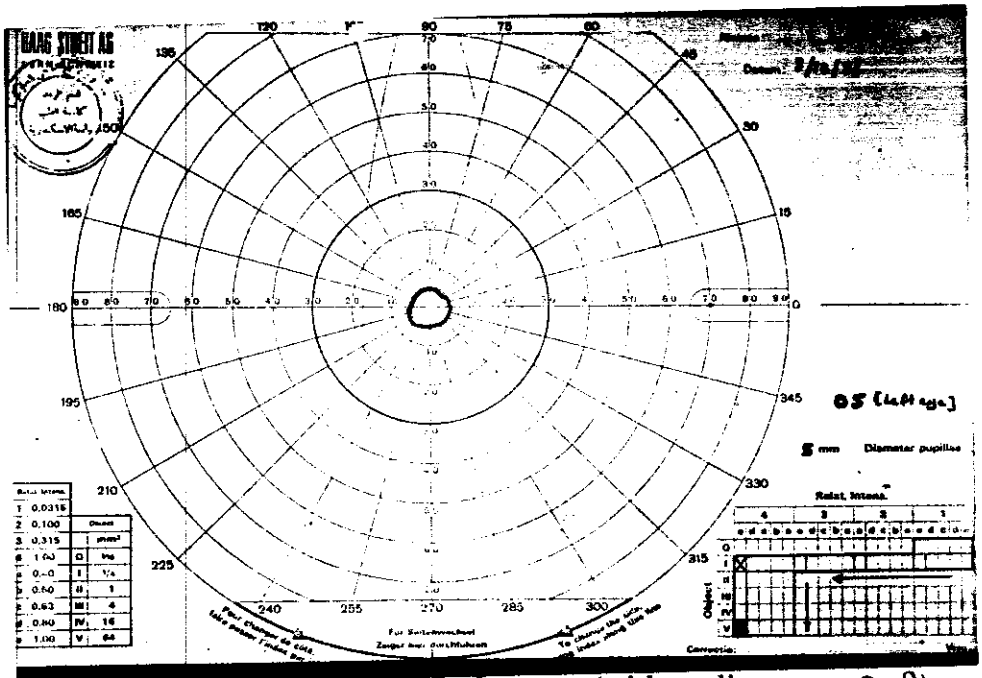

Fig.(15): Group III visual field changes (widest diameter <20°).

Table (XX): Prevalence of Posterior Subcapsular Cataract (PSC) Among 60 Eyes Having RP, by Genetic Type.

Posterior subcapsular cataract (PSC)	Genetic type					
	AD	AR	XL	S	UD	TOTAL
Clear lens	2 (50%)	16 (42.1%)	-	2 (14.3%)	-	20 (33.3%)
Cataract	2 (50%)	22 (57.9%)	2 (100%)	12 (85.7%)	2 (100%)	40 (66.7%)
Total	4 (100%)	38 (100%)	2 (100%)	14 (100%)	2 (100%)	60 (100%)

The distribution of cataract among the AR group (57.9%) and S group (85.7%) was not statistically significant, where Z = 1.3216. All cases with observed cataract showed posterior subcapsular cataract either alone or combined with other less frequent types.

In Table XXI, the prevalence of posterior subcapsular cataract (PSC) is indicated according to age group. Among the entire population, there was a clear tendency for PSC to develop with advancing age (Fig. 16), but even among the 26 eyes of those less than 32 years old, cataract was already present in 10 eyes (38.5%). Cataract was seen in 30 eyes (88.2%) of 34 eyes above 32 years of age. In other words, 80% of eyes with clear lens were aged 32 years or less in comparison to only 20% having more than 32 years. This difference was statistically significant where Z = 2.8558.

As regards the relation between prevalence of cataract and visual field changes (in 21 patients), table XXII and Fig. 17 show that 80% of patients in group I visual field had clear lens and the remaining 20% had cataract. In group II, visual field, 62.5% of the cases had cataract.

INTRAOCULAR PRESSURE IN RP:

The intraocular pressure was less than 22 mmHg in all patients included in this study. It was less than 16 mmHg in 85% of the eyes

Table (XXI): Prevalence of Posterior Subcapsular Cataract (PSC) Among 30 Patients (60 eyes) Having RP, by Age Group.

Posterior subcapsular cataract (PSC)	Age group				Total	
	< 32 years		> 32 years			
	Patients	Eyes	Patients	Eyes	Patients	Eyes
Clear lens	8 (61.5%)	16 (61.5%)	2 (11.8%)	4 (11.8%)	10 (33.3%)	20 (33.3%)
Cataract	5 (38.5%)	10 (38.5%)	15 (88.2%)	30 (88.2%)	20 (67.7%)	40 (67.7%)
Total	13 (100%)	26 (100%)	17 (100%)	34 (100%)	30 (100%)	60 (100%)

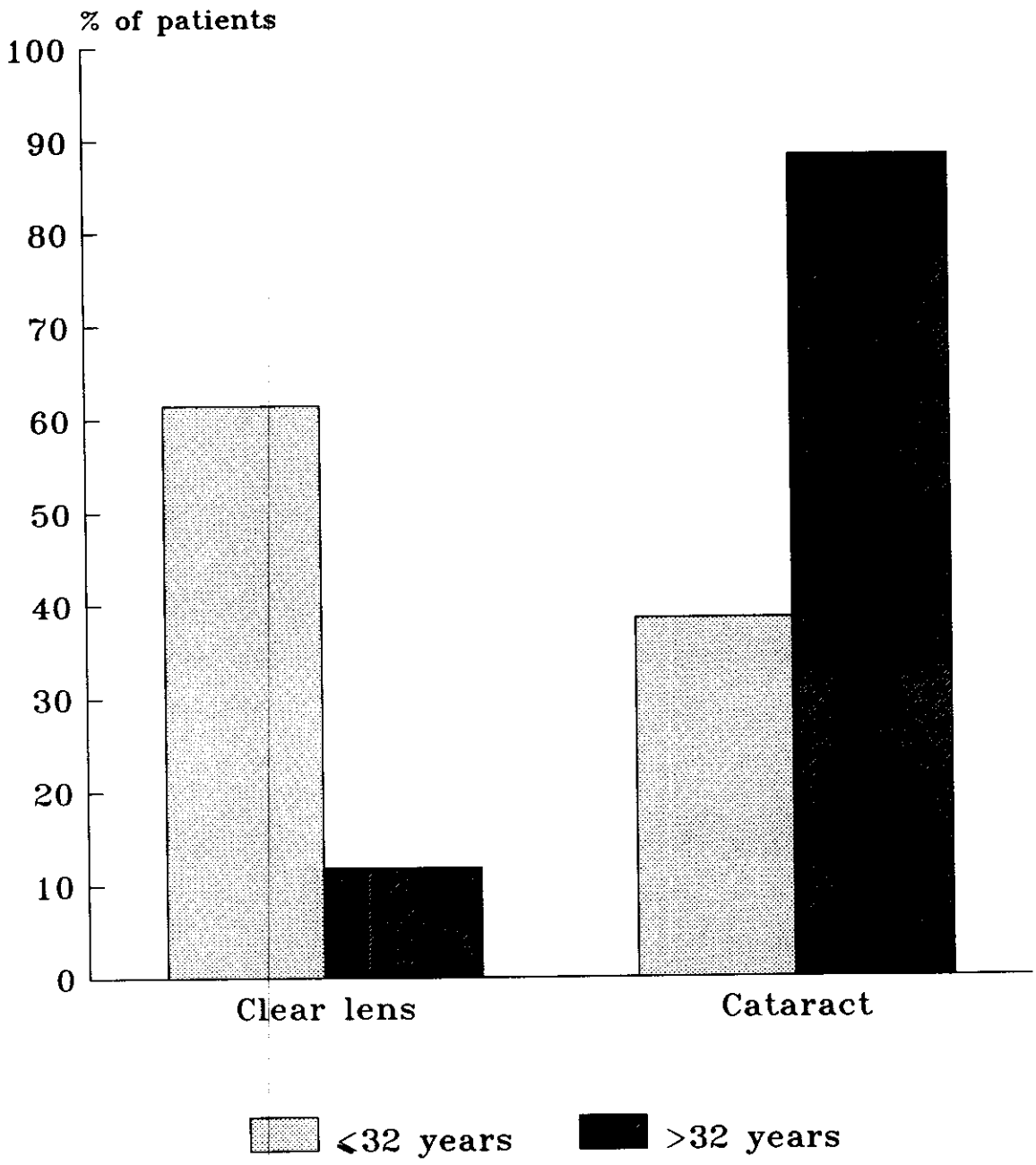

Fig.(16): Distribution of cataract according to age group.

Table (XXII): Prevalence of Posterior Subcapsular Cataract (PSC) Among 21 Patients Having RP by Visual Field Changes.

Field changes	Posterior subcapsular cataract (PSC)		Total
	Clear lens	Cataract	
Group I (>40°)	4 (40)	1 (9.1%)	5 (23.8%)
Group II (20°-40°)	3 (30%)	5 (45.5%)	8 (38.1%)
Group III (<20°)	3 (30%)	5 (45.5%)	8 (38.1%)
Total	10 (100%)	11 (100%)	21 (100%)
X	34.80	22.18	
"S"	19.52	14.16	

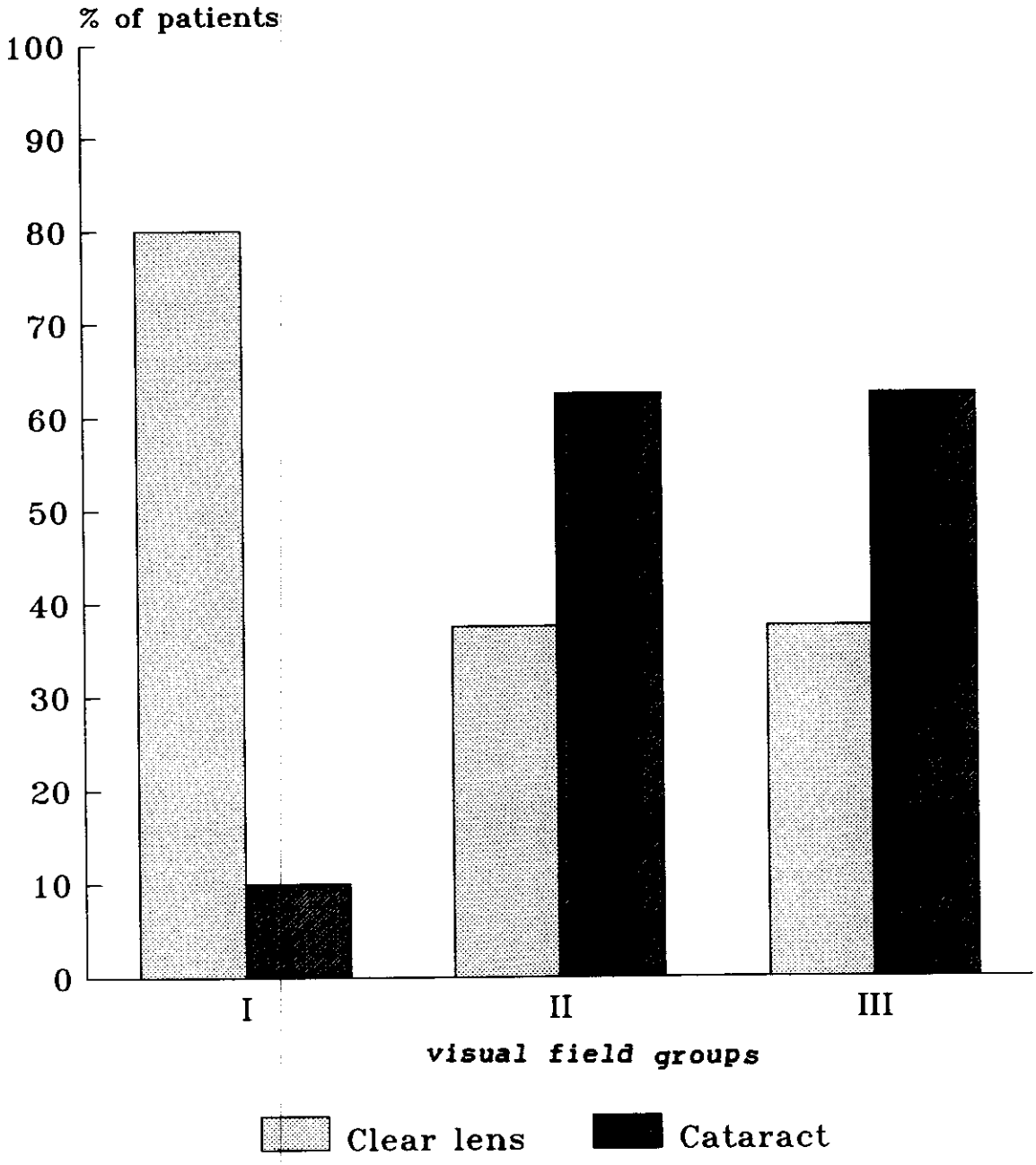

Fig.(17): Distribution of cataract according to visual field changes.

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(Table XXIII). The ocular tension ranged from 10-21 mmHg with a mean value of 13.22±2.77 mmHg.

OCULAR MOTILITY:

Concomitant exotropia was found in 2 patients (6.6%); all belonged to the AR group.

OPTIC DISC CHANGES:

Because "waxy pallor" of the optic disc is a hallmark of RP and because disc colour may, in part, reflect the vascular status of the fundus, eyes in this study were classified as showing pink (normal colour), creamy and waxy-appearing optic discs (Figs. 18-20). It is worth mentioning that some patients have a pink-appearing though not necessarily normal discs.

Table XXIV illustrates the distribution of optic disc colour in the different genetic types. Pink-appearing optic disc was present in 30% of eyes of RP patients and 42.1% of the eye of AR group. On the other hand, waxy-appearing optic disc was present in 23.3% of eyes of RP patients and 21.1% of eyes of AR group. As regards creamy-appearing disc, it was present in 46.7% of eyes of RP patients, 36.8% of eyes of AR group and 85.7% of eyes of S group. This was found to be statistically significant ($Z = 2.212$, $P < 0.05$).

Table (XXIII): Distribution of Intraocular Pressure Among a Series of 60 Eyes Having RP.

Intraocular pressure (mmHg)	Eyes	
	No.	%
10 - 15	51	85
16 - 21	9	15
Total	60	100

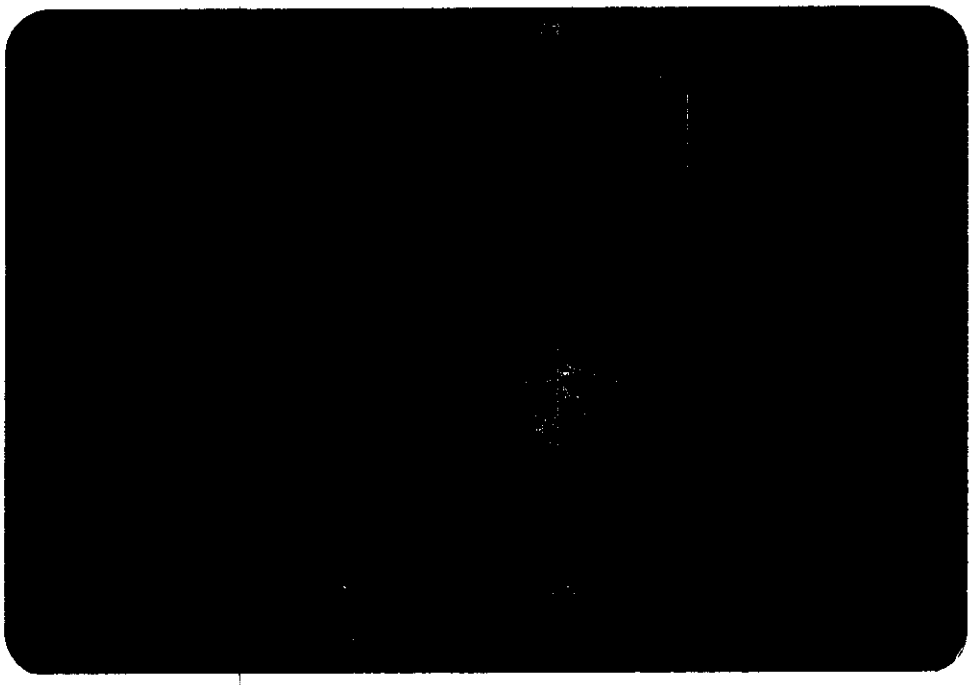

Fig.(18): Mild retinal vessel attenuation with pink-appearing optic disc.

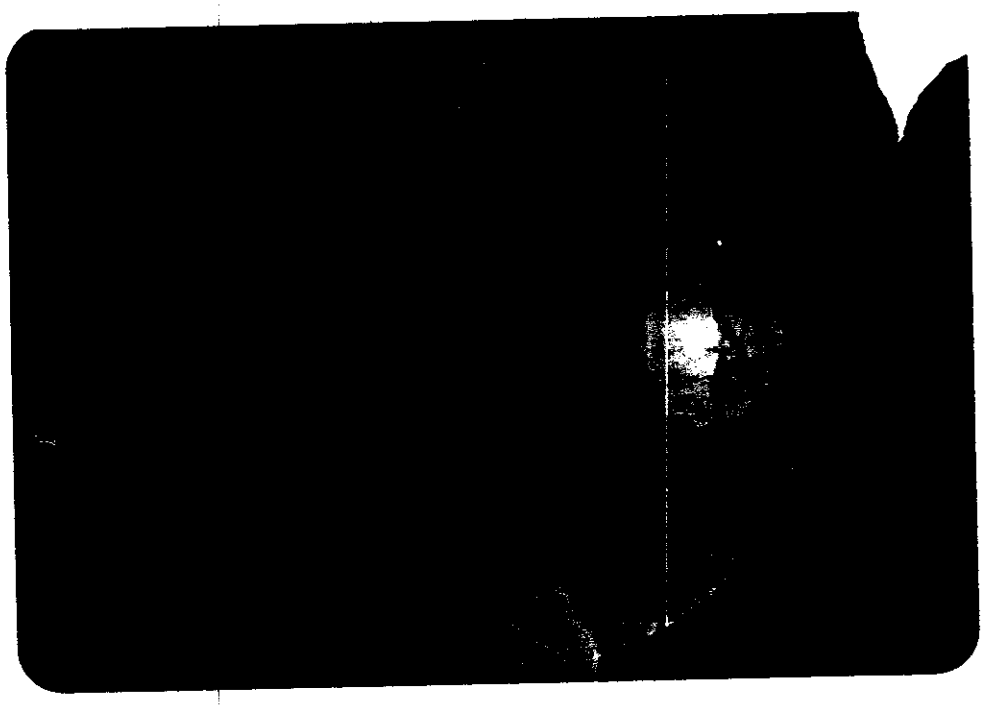

Fig.(19): Moderate retinal vessel attenuation with creamy-appearing optic disc.

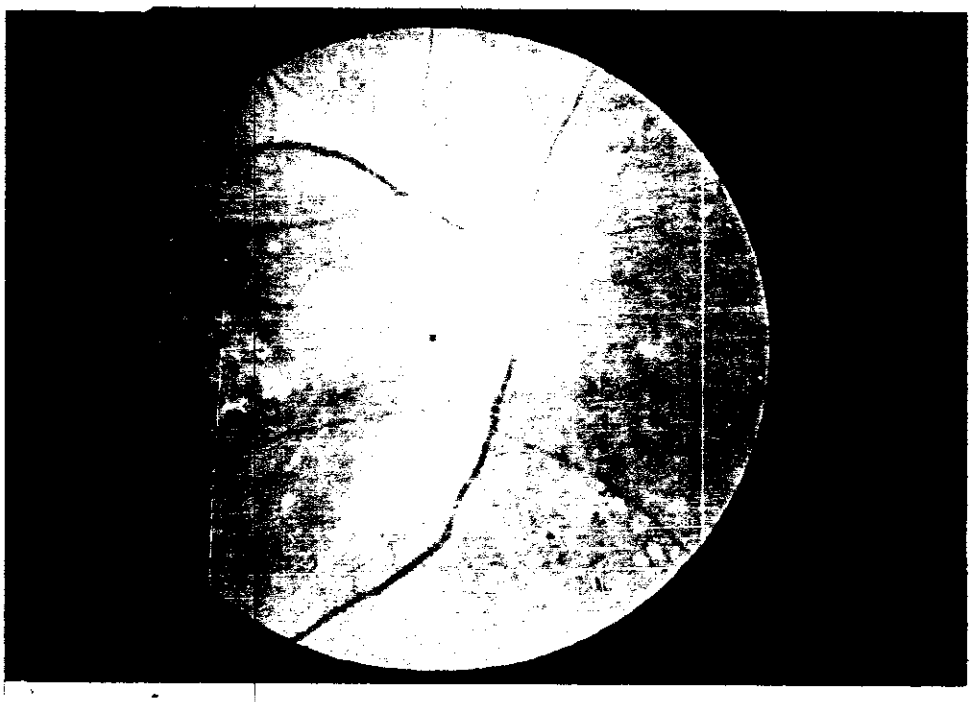

Fig.(20): Severe retinal vessel attenuation with waxy-appearing optic disc.

Table (XXIV): Distribution of Optic Disc Colour Among a Series of 60 Eyes Having RP, by Genetic Type.

Optic disc colour	Genetic type					
	AD	AR	XL	S	UD	TOTAL
Pink	2 (50%)	16 (42.1%)	-	-	-	18 (30%)
Creamy	2 (50%)	14 (36.8%)	-	12 (85.7%)	-	28 (46.7%)
Waxy	-	8 (21.1%)	2 (100%)	2 (14.3%)	2 (100%)	14 (23.3%)
Total	4 (100%)	38 (100%)	2 (100%)	14 (100%)	2 (100%)	60 (100%)

Table XXV demonstrates the relationship between optic disc colour and age group in RP patients. All RP patients with pink-appearing optic discs were less than or equal to 32 years old. In addition, 69.2% of patients aged less than or equal to 32 years had pink-appearing disc in comparison to 15.4% who had waxy-appearing disc. This difference was found to be statistically significant where $Z = 4.1004$, $P < 0.05$.

Table XXVI and Figures 21,22 illustrate the relationship between optic disc colour and visual field changes. It was shown that in patients with pink-appearing optic disc, 62.5% fitted group I visual field affection, whereas none of the patients belonged to group III visual field affection. This difference was statistically significant where $Z = 3.2656$, $P < 0.05$.

Also, there was a statistically significant difference between the incidence of pink-appearing disc on one hand (42.1%) and both creamy and waxy-appearing discs on the other hand (57.9%) in AR group where $Z = 2.0631$, $P < 0.05$.

RETINAL PIGMENTATION:

The density of retinal pigmentation was categorized into mild, moderate and severe (dense) according to Figs 23-25.

The presence of retinal pigmentation was a constant finding in all patients. Table XXVII shows that the distribution of severe retinal

Table (XXV): Distribution of Optic Disc Colour Among a Series of 30 Patients Having RP, by Age group.

Optic disc colour	Age group		Total
	⩽ 32 years	> 32 years	
Pink	9 (69.2%)	-	9 (30%)
Creamy	2 (15.4%)	12 (70.1%)	14 (46.71%)
Waxy	2 (15.4%)	5 (29.4%)	7 (23.3%)
Total	13 (100%)	17 (100%)	30 (100%)

Table (XXVI): Distribution of Optic Disc Colour Among a Series of 21 Patients Having RP, by Visual Field Changes.

Optic disc colour	Visual Field-Changes			Total
	Group I (>40°)	Group II (20°-40°)	Group III (<20°)	
Pink	5 (62.5%)	3 (37.5%)	-	8 (38.1%)
Creamy	-	5 (41.7%)	7 (58.3%)	12 (57.1%)
Waxy	-	-	1 (100%)	1 (100%)
Total	5 (23.8%)	8 (38.1%)	8 (38.1%)	21 (100%)

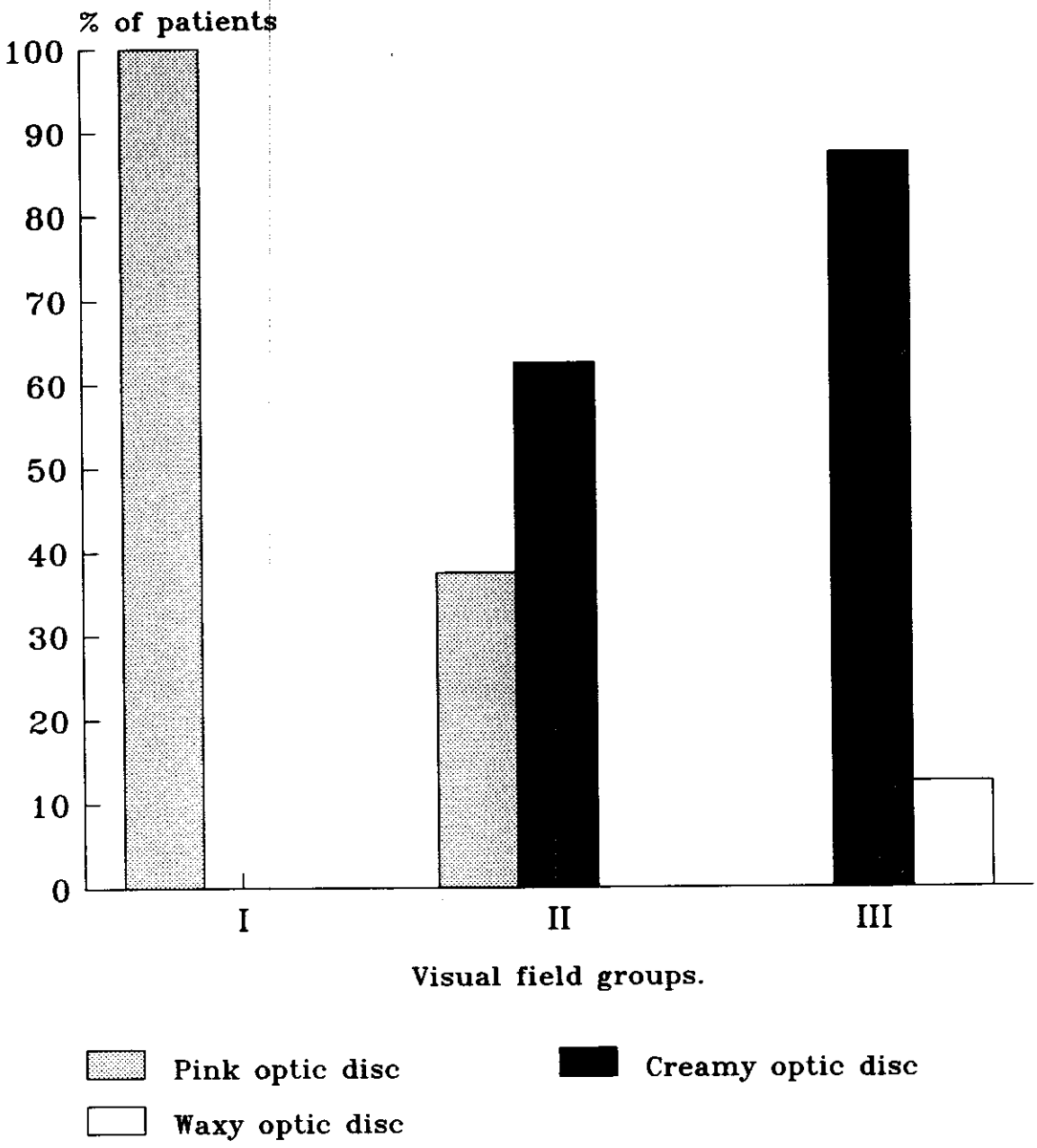

Fig.(21): Distribution of optic disc colour according to visual field changes.

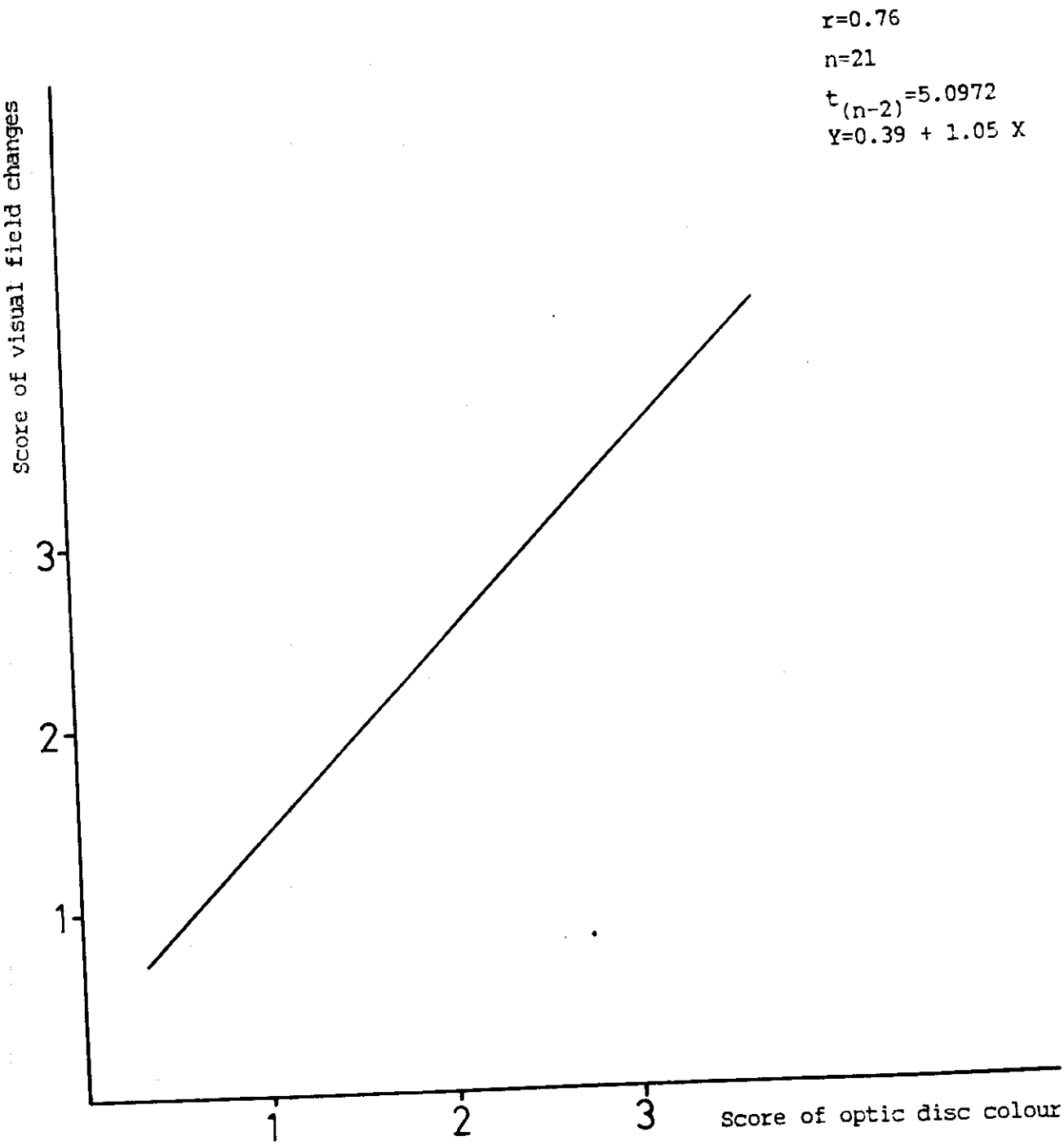

Fig.(22): Correlation between score of optic disc colour and score of visual field changes.

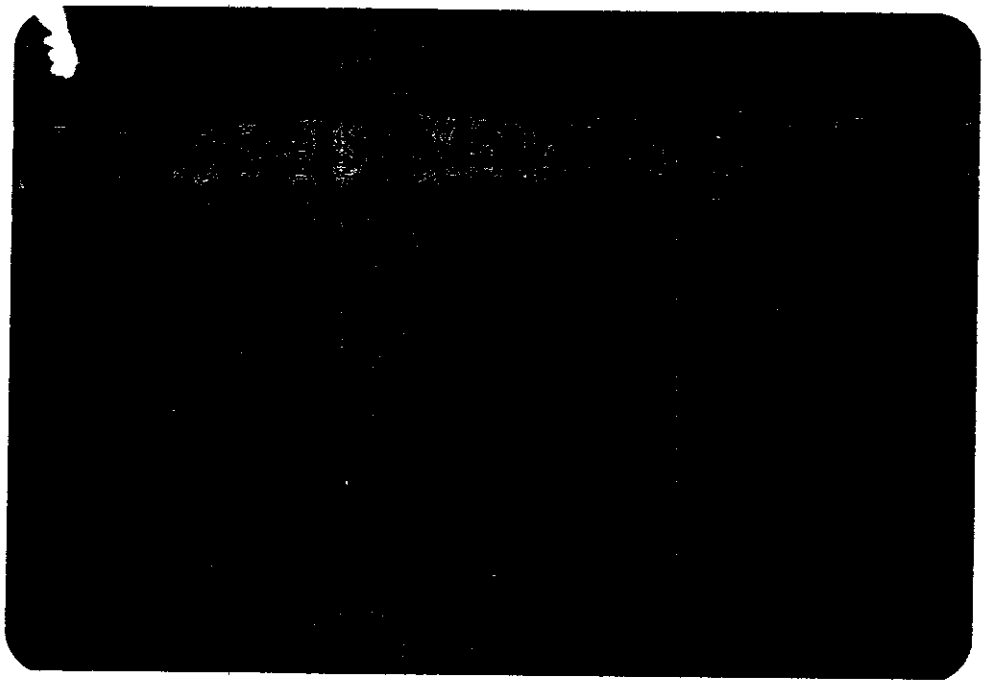

Fig.(23): Mild degree of retinal pigmentation.

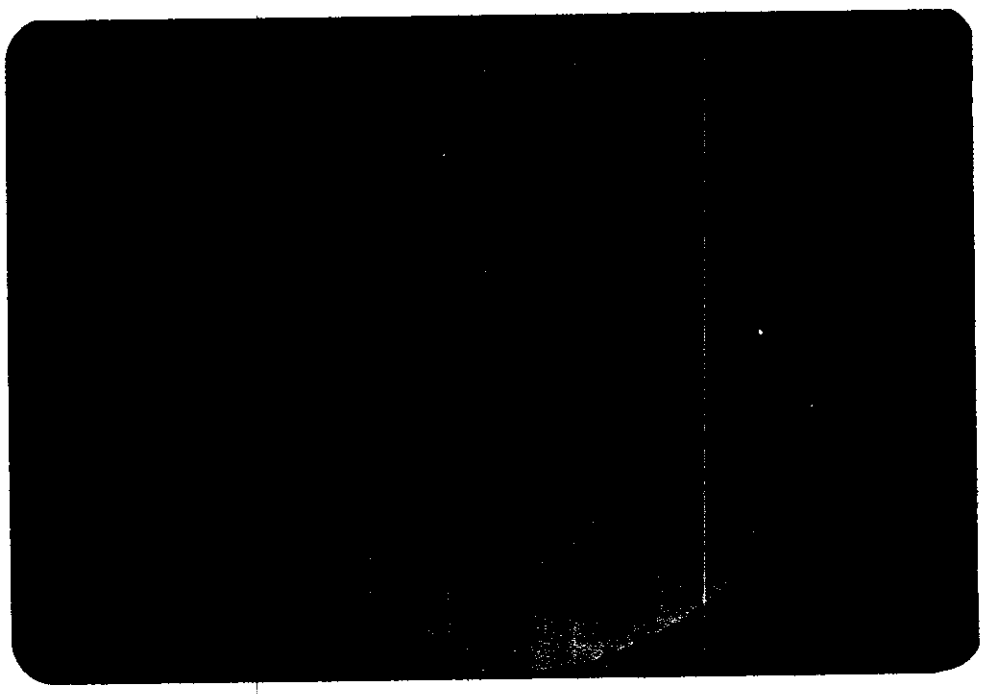

Fig.(24): Moderate degree of retinal pigmentation (Both bone corpuscles and rounded pigmentary spots are seen. Also pigments can be seen around retinal blood vessels).

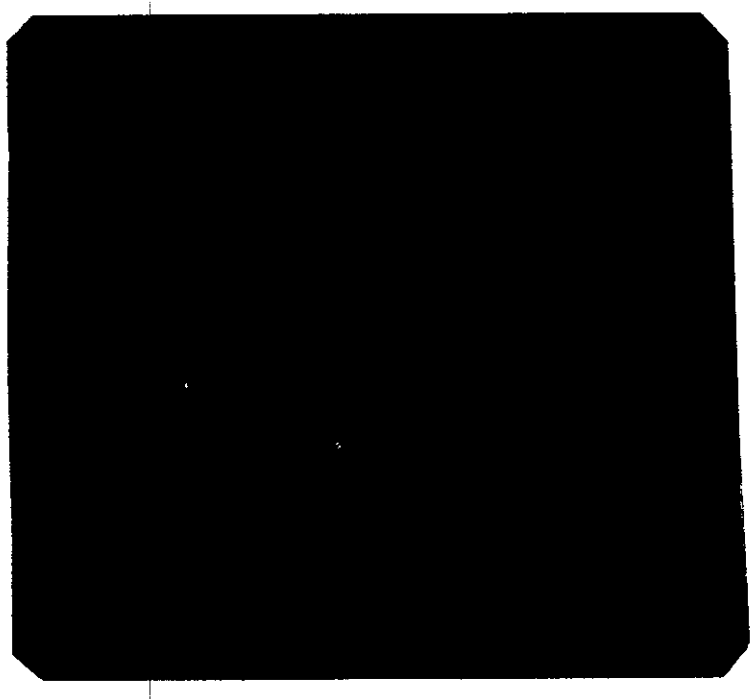

Fig.(25): Severe degree of retinal pigmentation (Patchy type).

Table (XXVII): Distribution of Retinal Pigmentation Density Among a Series of 60 Eyes Having RP, by Genetic Type.

Retinal pigmentation density	Genetic type					
	AD	AR	XL	S	UD	TOTAL
Mild	2 (50%)	16 (42.1%)	-	-	2 (100%)	20 (33.33%)
Moderate	2 (50%)	16 (42.1%)	-	6 (42.9%)	-	24 (40%)
Severe	-	6 (15.8%)	2 (100%)	8 (57.1%)	-	16 (26.7%)
Total	4 (100%)	38 (100%)	2 (100%)	14 (100%)	2 (100%)	60 (100%)

pigmentation was significantly more in the simplex type (85.7%) compared to the autosomal recessive type (15.8%) where $Z = 2.106$, $P < 0.05$. On the other hand, mild pigmentation was significantly more in the AR type (42%) compared to the S type where $Z = 2.0631$, $P < 0.05$. Also, the same table shows that the overall incidence of the mild, moderate and severe pigmentation in the RP patients was 33.3%, 80% and 26.7% respectively.

Table XXVIII illustrates the relationship between density of retinal pigmentation and age group of RP patients. Twenty per cent of patients with mild retinal pigmentation were >32 years old, whereas 87.5% of patients with severe retinal pigmentation were $\leq$32 years old. This difference was statistically significant, where $Z = 2.846$, $P < 0.05$.

Table XXIX and figure 26 demonstrate the relationship between the density of retinal pigmentation and optic disc colour. The incidence of mild retinal pigmentation was much higher in eyes with pink-appearing optic disc (80%) than in eyes with creamy and waxy discs (20%). Similarly, 88.9% of eyes with pink-appearing optic discs had mild pigmentation compared to 9.5% of eyes with creamy and waxy discs. These figures were found to be statistically significant where $r = 0.53$ ($t_{(n-2)} = 3.3072$, $P < 0.05$).

Table XXX and figure 27 show that in patients with group III visual field changes, 75% had severe retinal pigmentation whereas 12.5% had mild retinal pigmentation. Also, the distribution of severe retinal

Table (XXVIII): Distribution of Retinal Pigmentation Density Among a Series of 30 Patients Having RP, by Age group.

Retinal pigmentation density	Age-group		Total
	⩽ 32 years	> 32 years	
Mild	8 (61.5%)	2 (11.8%)	10 (33.3%)
Moderate	4 (30.8%)	8 (47.1%)	12 (40%)
Severe	1 (7.7%)	7 (41.1%)	8 (26.7%)
Total	13 (100%)	17 (100%)	30 (100%)

Table (XXIX): Distribution of Retinal Pigmentation Density Among a Series of 60 Eyes Having RP, by Optic Disc Colour.

Retinal pigmentation density	Optic disc colour			Total
	Pink	Creamy	Waxy	
Mild	16 (88.9%)	2 (7.1%)	2 (14.3%)	20 (33.3%)
Moderate	2 (11.1%)	14 (50%)	8 (57.1%)	24 (40%)
Severe	-	12 (42.9%)	4 (28.6%)	16 (26.7%)
Total	18 (100%)	28 (100%)	14 (100%)	60 (100%)

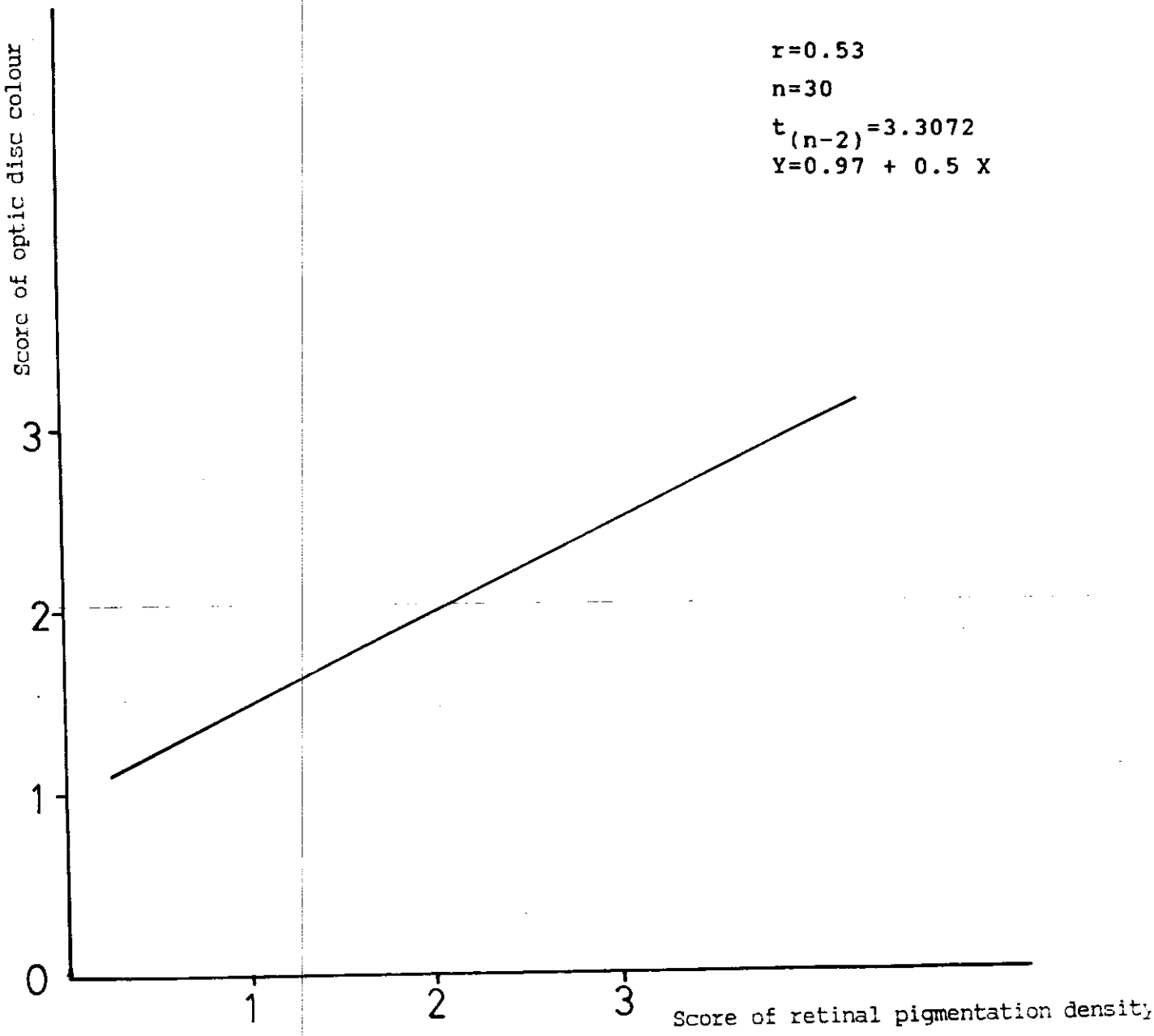

Fig.(26): Correlation between score of retinal pigmentation density and score of optic disc colour.

Table (XXX): Distribution of Retinal Pigmentation Density Among a Series of 21 Patients Having RP, by Visual Field Changes.

Retinal pigmentation density	Visual Field-Changes			Total
	Group I (>40°)	Group II (20°-40°)	Group III (<20°)	
Mild	3 (42.9%)	3 (42.9%)	1 (14.3%)	7 (33.3%)
Moderate	2 (33.3%)	3 (50%)	1 (16.7%)	6 (28.6%)
Severe	-	2 (25%)	6 (75%)	8 (38.1%)
Total	5 (23.8%)	8 (38.1%)	8 (38.1%)	21 (100%)

pigmentation was significantly more in patients with group III visual field affection (75%) compared to patients with grade I visual field affection (zero per cent), where $r = 0.57$ ($t_{(n-2)} = 3.0239$).

RETINAL VESSEL ATTENUATION:

The degree of retinal vessel attenuation was categorized into mild, moderate and severe according to Figs 18-20.

Attenuation of the retinal blood vessels was a constant finding in all patients (Table XXXI). However, 6.7% of the eyes involved showed a mild degree of vessel attenuation. Severe retinal vessel attenuation was present in 43.3% of the eyes of the RP patients and in 31.6% of the eyes of AR patients. The distribution of vessel attenuation showed no significant difference between the AR and S types of RP (mild, $Z = 0.619$, Moderate $Z = 1.5691$, Severe $Z = 1.8243$).

The incidence of severe retinal vessel attenuation was higher in patients > 32 years old (84.6%) than in patients $\leqslant 32$ years old (15.4%). This difference was found to be statistically significant where $Z = 2.5192$ (Table XXXII).

Table XXXIII and figure 28 demonstrate the relationship between the degree of retinal vessel attenuation and density of retinal pigmentation. The incidence of dense retinal pigmentation in eyes with

Table (XXXI): Distribution of Degree of Retinal Vessel Attenuation Among a Series of 60 Eyes Having RP, by Genetic Type.

Retinal vessel attenuation	Genetic type					
	AD	AR	XL	S	UD	TOTAL
Mild	2 (50%)	2 (5.3%)	-	-	-	4 (6.7%)
Moderate	2 (50%)	24 (63.1%)	-	4 (28.6%)	-	30 (50%)
Severe	-	12 (31.6%)	2 (100%)	10 (71.4%)	2 (100%)	26 (43.3%)
Total	4 (100%)	38 (100%)	2 (100%)	14 (100%)	2 (100%)	60 (100%)

Table (XXXII): Distribution of Degree of Retinal Vessel Attenuation Among a Series of 30 Patients Having RP, by Age group.

Retinal vessel attenuation	Age-group		Total
	≤ 32 years	> 32 years	
Mild	2 (15.4%)	-	2 (6.7%)
Moderate	9 (69.2%)	6 (35.3%)	15 (50%)
Severe	2 (15.4%)	11 (64.7%)	13 (43.3%)
Total	13 (100%)	17 (100%)	30 (100%)

Table (XXXIII): Degree of Retinal Vessel Attenuation In Relation to Retinal Pigmentation Density Among a Series of 60 Eyes Having RP.

Retinal vessel attenuation	Retinal pigmentation density			Total
	Mild	Moderate	Severe	
Mild	4 (20%)	-	-	4 (6.7%)
Moderate	14 (70%)	14 (58.3%)	2 (12.5%)	30 (50%)
Severe	2 (10%)	10 (41.7%)	14 (87.5%)	26 (43.3%)
Total	20 (100%)	24 (100%)	16 (100%)	60 (100%)

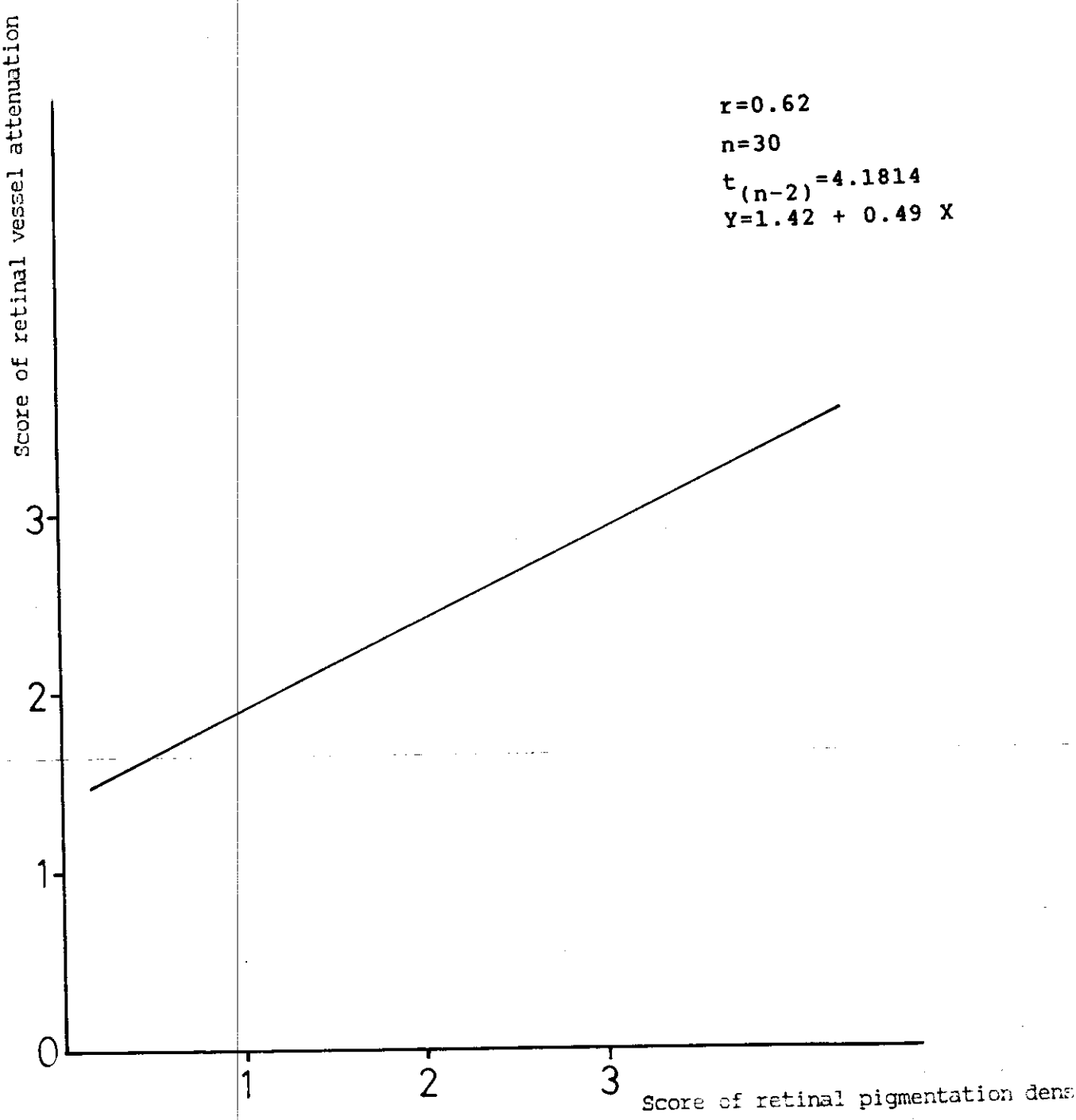

Fig.(28): Correlation between score of retinal vessel attenuation and score of retinal pigmentation density.

Table (XXXIV): Degree of Retinal Vessel Attenuation In Relation to Optic Disc Colour Among a Series of 60 Eyes Having RP.

Retinal vessel attenuation	Optic disc colour			Total
	Pink	Creamy	Waxy	
Mild	4 (20%)	-	-	4 (6.7%)
Moderate	16 (80%)	12 (46.2%)	2 (14.3%)	30 (50%)
Severe	-	14 (53.8%)	12 (85.7%)	26 (43.3%)
Total	20 (100%)	26 (100%)	14 (100%)	60 (100%)

Table (XXXV): Degree of Retinal Vessel Attenuation in Relation to Visual Field Changes Among a Series of 21 Patients Having RP.

Retinal vessel attenuation	Visual Field Changes			Total
	Group I (>40°)	Group II (20°-40°)	Group III (<20°)	
Mild	-	1 (12.5%)	-	1 (4.8%)
Moderate	5 (100%)	6 (75%)	2 (25%)	13 (61.9%)
Severe	-	1 (12.5%)	6 (75%)	7 (33.3%)
Total	5 (23.8%)	8 (38.1%)	8 (38.1%)	21 (100%)

DISTRIBUTION OF PATIENTS ACCORDING TO SHEATHING OF RETINAL VESSELS: (Fig 31)

Table XXXVI demonstrates that sheathing of the retinal vessels was found in 23.3% Of eyes of RP patients, 26.3% Of eyes of AR group and 28.6% Of eyes of s group. Table XXXVII shows that 85.7% of eyes with sheathing of retinal vessels belonged to patients above 32 years of age. On the other hand, 92.3% of patients ≤ 32 years old did not show sheathing of retinal vessels.Table XXXVIII illustrates the relationship between sheathing of retinal vessels and retinal pigmentation density. The table shows that 37.5% of eyes with severe retinal pigmentation had sheathing in comparison to only 10% of eyes with mild retinal pigmentation.

RP patients having sheathing of retinal vessels were characterized by poor visual field and none of them belonged to group I visual field changes. As regards visual acuity, sheathing was present in eyes with affected vision (low and blind).

Macular changes:

The common macular changes encountered in this study are shown in figures 32-44. Macular changes were evaluated by both ophthalmoscopy and fluorescein angiography. Table XXXIX shows the incidence of the macular changes in RP and in its different genetic types. There was one

Fig.(31a): Fundus photograph of the right eye of a 30-year-old male with AR type of RP showing sheathing and pigmentation along the retinal vessles. Also a thin white rim encompassing the optic disc can be seen.

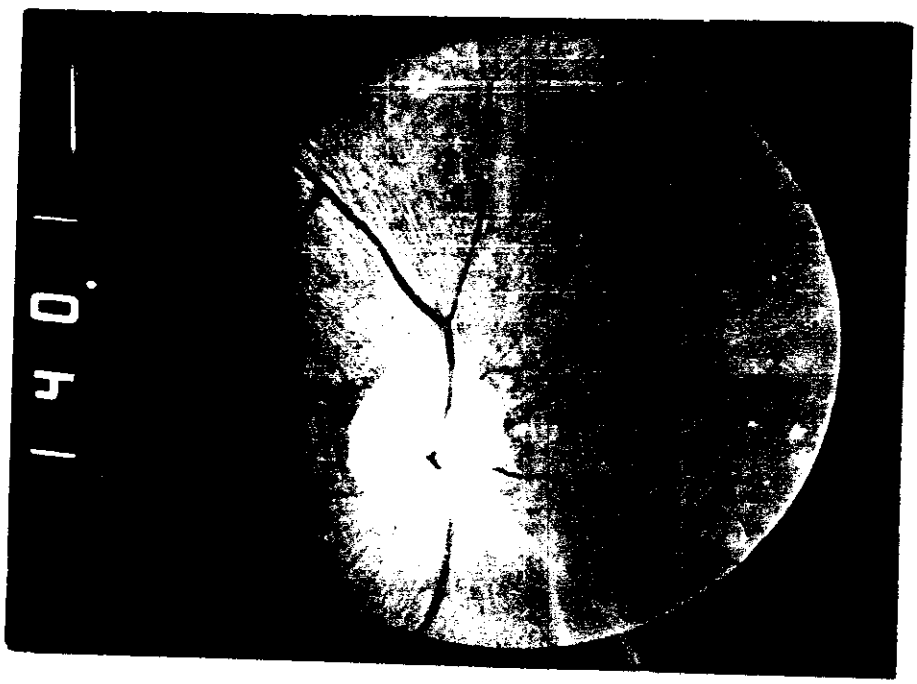

Fig.(31b): Red free photograph of the same eye demonstrating an area of nerve fibre layer drop out superior to the optic disc. Segmentation of the blood column inside the retinal blood vessels can also be seen.

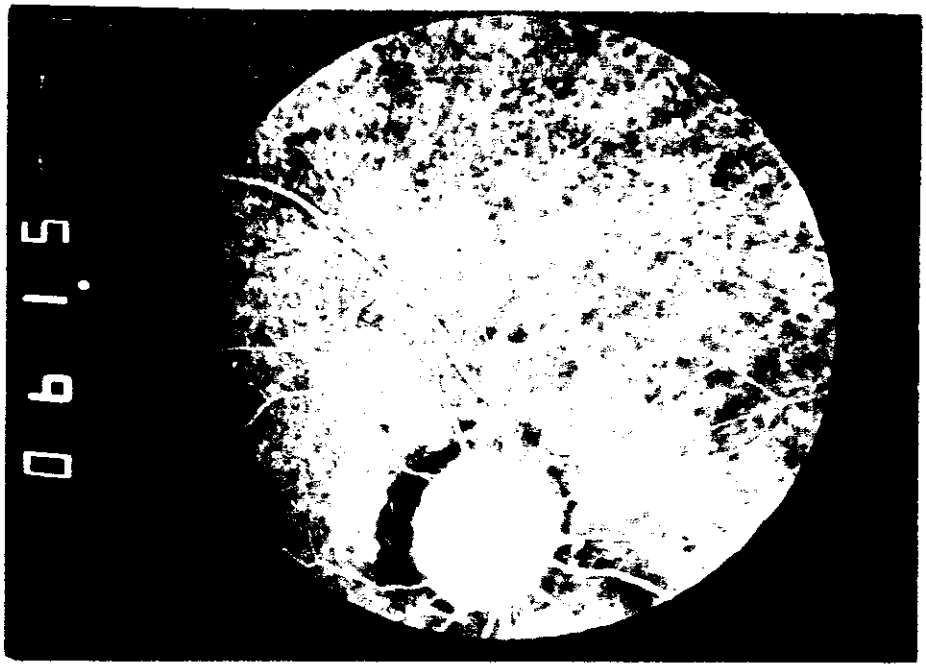

Fig.(31c): Fluorescein angiogram, late venous phase, showing that the retinal blood flow is not blocked by the retinal pigments and sheathing along the retinal vessels. The angiogram also highlights the peripapillary changes.

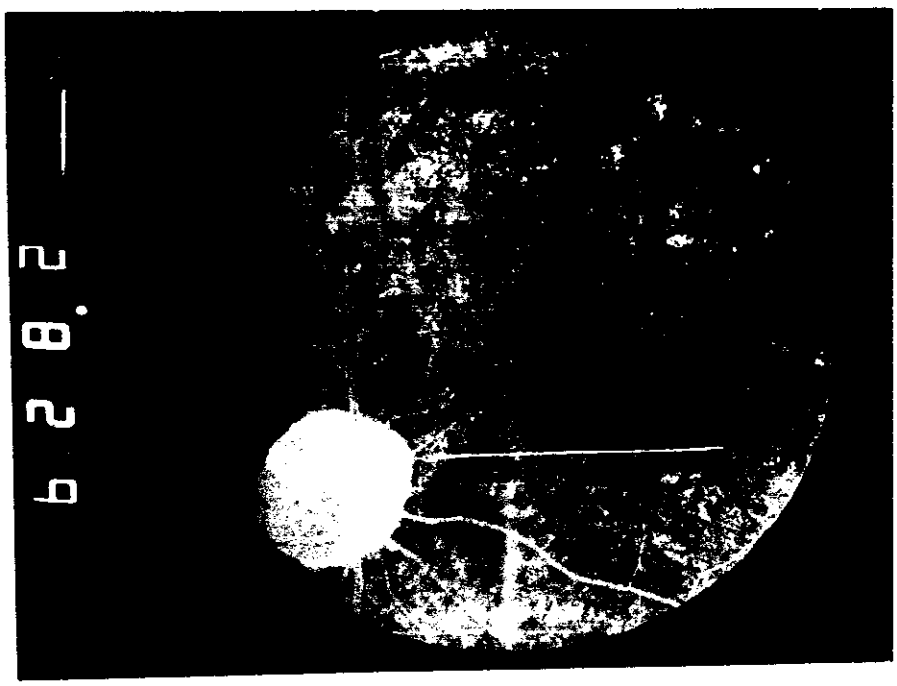

Fig.(31d): Fluorescein angiogram, late phase, demonstrating the staining (hyperfluorescence) of the thin rim encompassing the optic nervehead.

Table (XXXVI): Prevalence of Sheathing of Retinal Vessels Among a Series of 60 Eyes Having RP, by Genetic Type.

Sheathing of retinal vessels	Genetic type					
	AD	AR	XL	S	UD	TOTAL
Present	-	10 (26.3%)	-	4 (28.6%)	-	14 (23.3%)
Absent	4 (100%)	28 (73.7%)	-	10 (71.4%)	2 (100%)	46 (76.7%)
Total	4 (100%)	38 (100%)	2 (100%)	14 (100%)	2 (100%)	60 (100%)

Table (XXXVII): Distribution of Sheathing of Retinal Vessels Among a Series of 30 Patients Having RP, by Age group.

Sheathing of retinal vessels	Age-group		Total
	≤ 32 years	> 32 years	
Present	1 (7.7%)	6 (35.3%)	7 (23.3%)
Absent	12 (92.3%)	11 (64.7%)	23 (76.7%)
Total	13 (100%)	17 (100%)	30 (100%)

Table (XXXVIII): Sheathing of Retinal Vessels In Relation to Retinal Pigmentation Density Among a Series of 60 Eyes Having RP.

Sheating of retinal vessels	Retinal pigmentation density			Total
	Mild	Moderate	Severe	
Present	2 (10%)	6 (25%)	6 (37.5%)	14 (23.3%)
Absent	18 (90%)	18 (75%)	10 (42.5%)	46 (76.7%)
Total	20 (100%)	24 (100%)	16 (100%)	60 (100%)

Table (XXXIX): Macular Changes in the Different Genetic Types Among a Series of 60 Eyes Having RP.

Genetic types \ Macular changes	Normal	Enlarged hypofluor-escence	Atrophy	Bull's eye pattern	Pigmentation	Preretinal membrane (striate change	Macular oedema	Absent foveal reflex
AD (4)	-	2 (3.3%)	2 (3.3%)	-	-	-	-	4 (6.6%)
AR (38)	2 (3.3%)	24 (40%)	18 (30%)	2 (3.3%)	4 (6.6%)	14 (23.3%)	4 (6.6%)	30 (50%)
XL (2)	-	-	2 (3.3%)	-	2 (3.3%)	-	-	2 (3.3%)
S (14)	-	8 (13.3%)	8 (13.3%)	2 (3.3%)	2 (3.3%)	-	-	12 (20%)
UG (2)	-	-	2 (3.3%)	-	-	-	-	2 (3.3%)
Total (60)	2 (3.3%)	34 (56.7)	32 (53.3%)	4 (6.6%)	8 (13.3%)	14 (23.3%)	4 (6.6%)	50 (83.3%)

* Percent is from total number of eyes.

or more macular changes in 96.7% of studied eyes. The most prevalent macular change was absence of foveal reflex (83.3%). Enlarged macular hypofluorescense (56.7%) and macular atrophy (53.3%) were the next most frequently seen abnormalities. In all cases with enlarged macular hypofluorescense, the macular capillary bed was not clear. Macular atrophy (53.3%) was best diagnosed on angiographic basis in the form of window defect and/or staining of the RPE. Beside the RPE degeneration and atrophy, macular pigmentation was sometimes present (13.3%). Bull's eye appearance (6.6%) was evident clinically as a halo of pallor at the pigment epithelial level which showed a circular window defect on fluorescein angiography. A striate change (23.3%); cellophane-like, irregular light reflex was present and associated with fine corrugations of inner sensory retina that assumed parallel radial or whort-like configurations. In some, only the macula was affected; in others, the abnormality extended to the major temporal vascular arcades and the disc margin. Macular oedema, either in the form of diffuse or cystoid pattern was present in 6.6% of eyes.

Other macular changes included, tapetal reflex in 6.6% of eyes; 3.3% of eyes showed few dot macular haemorrhages (history of diabetes mellitus and telangiectatic capillary bed occurred in 3.3% of eyes (which also showed fluorescein leakage). Only 3.3 of eyes were categorized as normal.