AIM OF THE WORK

AIM OF THE WORK

The aim of the work was the early detection and study of the prevalence of congenital and developmental eyelid anomalies and the availability of treatment for such cases in patients attending Alexandria Main University Pediatric Ophthalmology Outpatient Clinic.

SUBJECTS

SUBJECTS

Screening of all the patients attending Alexandria Main University Pediatric Ophthalmology Outpatient Clinic under age of 15 years during a period of six months starting from December 2013.

METHODS

METHODS

In this study we assessed the prevalence, relative frequency and nature of congenital and developmental eyelid malformations among infants and children attending Alexandria Main University Pediatric Ophthalmology Outpatient Clinic.

Upon presentation every infant and child was subjected to:

➢ History taking from parents including

- Demographic data as regards name, age, gender and consanguinity.
- Family history for the same or other congenital anomalies.
- Referral of the discovered cases to the oculoplastic unit for further assessment and treatment.

➢ Ophthalmic examination with emphasis upon

- Careful external examination along with palpation of the eyelids and the orbital rim.
- Evaluation of any clinical evidence of relative proptosis or enophthalmos in each eye.
- Inspection of eyebrows at baseline was done, since patient may compensate by lifting eyebrows with the frontalis muscle.
- The patient's head posture was noted; document chin-up position.
- Visual acuity and cycloplegic refraction were done. This helped us to identify refractive errors and amblyopia. In infants, we noted if baby could fixate, maintain, and follow light.
- Pupillary examination was done. Pupillary size and iris color differences between the two eyes were examined to rule out Horner's syndrome.
- Extraocular muscle movement was evaluated with a note of diplopia (in chronic progressive external ophthalmoplegia, there may be extraocular muscle weakness together with ptosis).
- Dilated fundus examination ruled out any posterior segment pathology, such as abnormal retinal pigmentation seen in Kearn Sayre's syndrome.

➢ Eyelid measurements

- Quantifying the severity of ptosis, various eyelid measurements were taken with the face held in the frontal plane and with frontalis muscle relaxed.
- Neutralizing frontalis contracture by holding the brows in their relaxed position.
- If the droopy lid obscured the lid margin, we lifted the brows to observe the lid level for measurement.
- ***Palpebrae fissure height (PF)*** is the distance between the upper and lower eyelid margins at the axis of the pupil. Normal measurement is 9 to 12 mm.
- ***Marginal reflex distance (MRD)*** is the distance between the central corneal light reflex and upper eyelid margin with eyes in primary position. Normal MRD is 4-5 mm.

- ***Levator function*** is the distance the eyelid travels from downgaze to upgaze while the frontalis muscle is held inactive at the brow. The amount of lid elevation was recorded in millimeters (mm). Classification of levator function:
  Poor: 0 – 5 mm lid elevation
  Fair: 6 – 11 mm lid elevation
  Good: > 12 mm lid elevation

- ***Upper eyelid crease*** position is the distance from the upper eyelid crease to the upper eyelid margin. It is normally 7-8 mm in males and 9-10 mm in females.

➢ Ancillary testing

1. Prostagmine test was done whenever myasthenia gravis was suspected.
2. Serial external photographs of the eyes and the face were included in the patient's record for documentation, especially in children, when direct measurements were difficult.
3. Schirmer's test to evaluate tear function and fluorescein test for tear break-up time.
4. Jaw winking phenomenon.

RESULTS

RESULTS

The present study aimed to provide data about the prevalence of congenital eye anomalies in children attending Alexandria Main University Ophthalmology outpatients' clinic during a period of six months starting from December 2013.

A total of 9484 patients were examined for congenital and developmental eyelid anomalies (after taking parent's consent). Ninety nine eyes of eighty patients were found to have a congenital or developmental eyelid anomaly.

Patients once examined were not re-included in our study.

Descriptive statistics of the examined cases (n=9484)

Screening was done for 9484 children attending the Alexandria Main University Pediatric outpatient clinic during the period of December 2013 to May 2014 for congenital and developmental eyelid anomaly. The age ranged from 2 weeks to less than 15 year with a mean of 3.63 SD ±3.1. The 9484 child examined were studied according to their age distribution and gender among study's 6 month duration (Table 1)

Table (1): Showing descriptive statistics of the examined cases (n=9484).

	December (n=1490)	January (n=1685)	February (n=1590)	March (n=1734)	April (n=1042)	May (n=1943)
Age distribution						
<1year	450	500	475	535	441	843
1year -<5years	500	585	525	590	391	595
5years-<10years	400	362	439	411	161	405
10 years-<15 year	140	238	151	198	49	100
Gender						
Male	811	975	833	911	607	1141
Female	679	710	757	823	435	802

The highest prevalence of congenital and developmental eyelid anomalies among 9484 patients examined was ptosis with thirty nine patients affected (0.4%) of total examined cases followed by internal and external angular dermoids in fifteen cases (0.16%). (Table2)

Table (2): Showing prevalence of different eyelid anomalies among examined cases (n=9484).

Clinical characteristics	Examined cases (n=9484)	
	No.	%
Diagnosis		
Ptosis	39	0.4
Coloboma	12	0.12
Int. and Ext. angular Dermoid	15	0.16
Hemangioma	1	0.01
Symblepharon with NLDO	2	0.02
Kohn Romano	2	0.02
Lagophthalmos	1	0.01
Distichiasis	1	0.01
Dermolipoma	6	0.06
Telecanthus	1	0.01
Total	80	

Descriptive statistics of the affected cases (n=80)

Screening done on 9484 children attending the Alexandria Main University Pediatric outpatient clinic revealed eighty cases with positive clinical criteria for congenital and developmental eyelid anomaly. The age ranged from 3 months to 13 years with a mean of 4.0 SD ±3.6. (Table 3)

Table (3): Showing age among affected cases (n=80).

	Cases	Minimum	Maximum	Mean	Std. Deviation
Age (year)	80	0.25	13	4.0	±3.6

Table 4 showing the prevalence of congenital and developmental eyelid anomalies among children examined distribution in the period of December 2013 to May 2014 in Alexandria Main University Pediatric Outpatients' clinic. (Table 4)

Table (4): Showing prevalence among the affected cases (n=80).

Month	attendants	Cases	Prevalence/1000 attendants
December	1490	10	7
January	1685	19	11
February	1590	14	9
March	1734	11	6
April	1042	13	12
May	1943	13	7
Total	**9484**	**80**	**8**

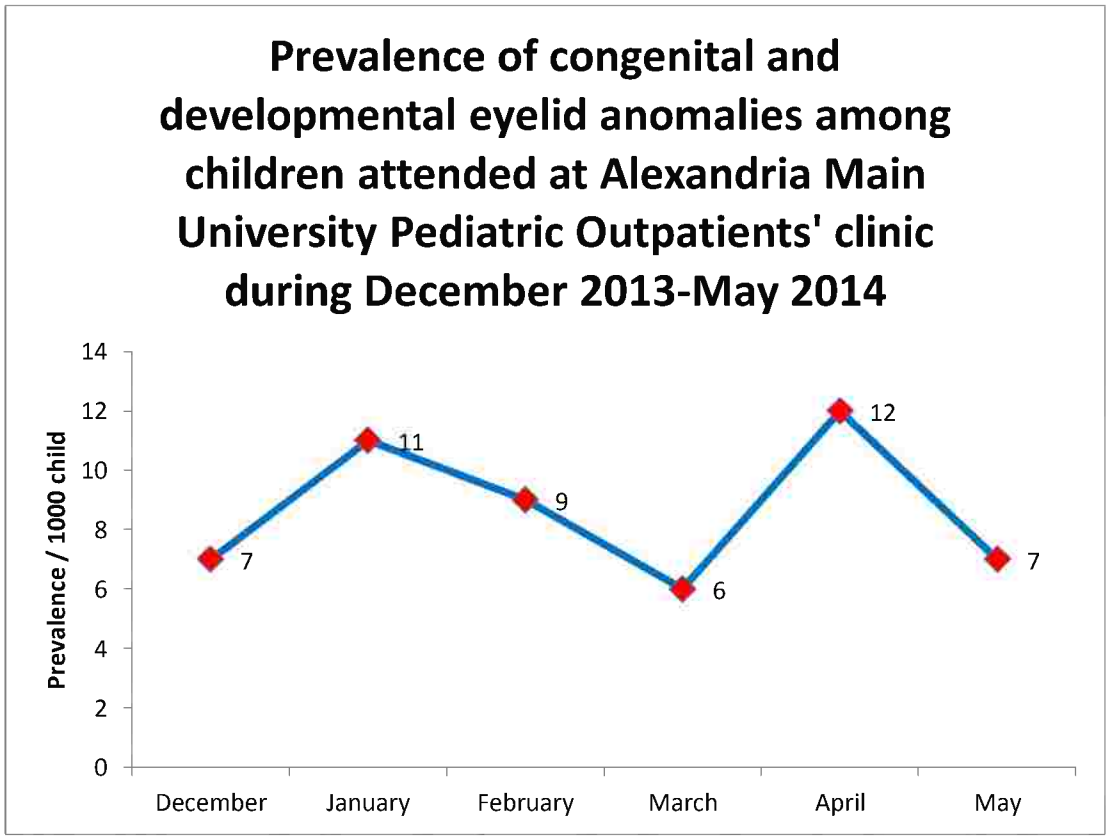

Figure (10): Showing prevalence of the affected cases (n=80).

Descriptive studies were done for 80 cases with positive eyelid anomalies and it revealed significant P-value for the age group one year to less than five years for the prevalence of congenital and developmental eyelid anomalies and there were no significant P-value regarding the gender for any of the total age groups studied. The study included 80 cases; 24 (30%) aged less than one year, 28 (35%) aged 1 to less than 5 years, 21 (26.25%) aged 5 to less than 10 years, 7 (8.75%) aged 10-15 year. The studied 80 cases included 44 male (55%) and 36 female (45%). (Table 5)

Table (5): Showing descriptive results of the affected cases (n=80).

	December (n=10)	**January (n=19)**	**February (n=14)**	**March (n=11)**	**April (n=13)**	**May (n=13)**	**p-value Test value**
Age distribution							
<1year	3	6	4	3	4	4	p=0.063 t=1.879
1year -<5years	4	6	5	4	5	4	p=0.007* t=2.776
5years-<10years	3	5	4	3	3	3	p=0.966 t=0.043
10 years-<15 year	0	2	1	1	1	2	p=0.725 t=0.352
Gender							
Male	6	11	8	7	8	4	p=0.536 t=0.62
Female	4	8	6	4	5	9	p=0.579 t=0.557

t: t-test *Significant at p-value ≤ 0.05

Among the 80 cases examined, 26 Right eyes (32.5%), 35 Left eyes (43.75%) and 19 bilaterally affected (23.75%) respectively. (Table 6)

Table (6): Showing laterality among the studied cases. (n=80)

Clinical characteristics	Studied cases (n=80)	
	No.	%
Laterality		
Right	26	32.5
Left	35	43.75
Bilateral	19	23.75

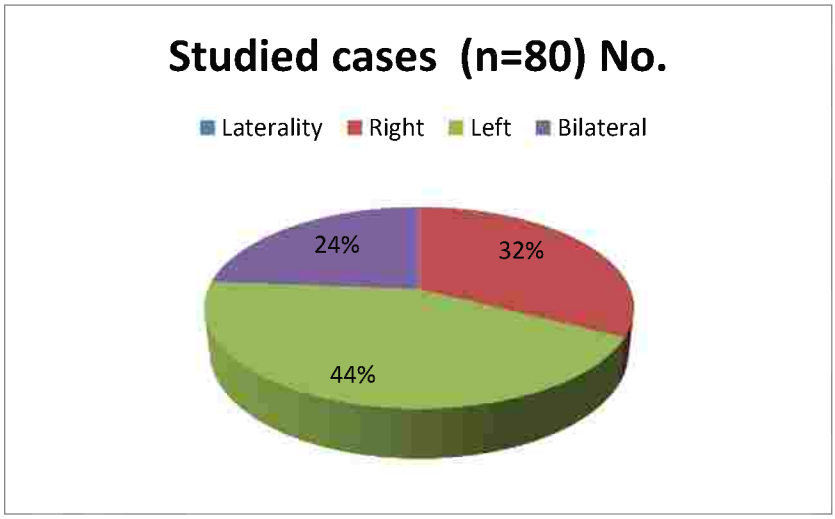

Figure (11): Showing laterality among the studied cases. (n=80)

Clinical characteristics of the 80 affected cases with congenital and developmental eyelid anomalies were assessed. Fundus examination was done (under general anaesthesia for 73 of the cases while as it was done without anaesthesia to 7 cases) showed normal picture except for one case (1.25%) found to have pale optic nerve head which was irrelevant to the acquired ptotic pathology.

Regarding the visual acuity, fix and follow could be assessed in 11 cases of the affected cases having congenital or developmental eyelid anomalies. They all could fix and follow. LEA charts was used with 39 cases. Visual acuity was measured in 30 cases using Snellen charts.

As for the refraction, 80 affected cases having congenital and developmental eyelid anomalies had their refraction measured. The refraction ranged from (-2.0) to (+4.25) diopters.

Two cases were found to have amblyopia in one eye, both had ptosis and their age was 3 and 5 years respectively.

Anterior segment examination was done on all cases having congenital and developmental eyelid anomalies. One eye with anterior chamber (AC) dysgenesis was found in a patient having dermolipoma in his right eye aging 1.4 years. (Table 7)

Table (7): Showing clinical characteristics among the affected cases. (n=80)

Clinical characteristics	Affected cases (n=80)	
	No.	%
Fundus pathology		
Present	1	1.25
Absent	79	98.75
Visual acuity		
Fix and follow	11	13.75
LEA chart	39	48.75
Snellen chart	30	37.5
Anterior Segment pathology		
Present	1	1.25
Absent	79	98.75

Correlations of different eyelid anomalies (n=99)

Eighty patients with congenital and developmental eyelid anomalies had 99 eyes affected; 26 (26.3%) aged less than one year, 36 (36.4%) aged 1 to less than 5years, 29 (29.3%) aged 5 to less than 10 years, 8 (8%) aged 10-15 years. There is a significant prevalence of ptosis in children aged 5 to less than 15 years while there is a significant prevalence of coloboma in children aged less than 5 years. (Table 8)

Table (8): Showing correlation between age groups and affected eyes. (n=99)

Eyelid anomalies	Affected eyes (n=99)				Significance
	Less than 5 (n=62)		5 years or more (n=37)		
	No.	%	No.	%	
Ptosis	24	38.7	30	81.1	**X^2=6.98 P=0.008***
Colobomas	11	17.7	1	2.7	**^{FE}P=0.029***
Int&Ext. A.Dermoid	11	17.7	4	10.8	X^2=0.87 P=0.352
Hemangioma	1	1.6	0	0.0	^{FE}P=1.0
Symblepharon	2	3.2	0	0.0	^{FE}P=0.527
Kohn Romano	4	6.5	0	0.0	^{FE}P=0.293
Lagophthalmos	1	1.6	0	0.0	^{FE}P=1.0
Distiachiasis	0	0.0	2	5.4	^{FE}P=0.137
Dermolipoma	6	9.7	0	0.0	^{FE}P=0.081
Telecanthus	2	3.2	0	0.0	^{FE}P=0.527

X^2: Chi-Square test ^{FE}P: Fisher's Exact test *significant at P≤0.05

Distribution of the ninety nine eyes with congenital and developmental eyelid anomalies according to gender showed that a total of 53 eyes were male (53.5%) and 46 eyes were female (46.5%). There was a significant prevalence of colobomas among females and dermolipoma among males. (Table 9)

Table (9): Showing gender distribution among eyes with detected eyelid anomalies.

Eyelid anomalies	Studied eyes (n=99)				Significance
	Male (n=53)		Female (n=46)		
	No.	%	No.	%	
Ptosis	32	60.4	22	47.8	X^2=1.56 P=0.211
Colobomas	1	1.9	11	23.9	**X^2=11.22 P=0.0008***
Int&Ext. A.Dermoid	9	17.0	6	13.0	X^2=0.3 P=0.586
Hemangioma	0	0.0	1	2.2	^{FE}P=1.0
Symblepharon	0	0.0	2	4.3	^{FE}P=0.214
Kohn Romano	2	3.8	2	4.3	^{FE}P=1.0
Lagophthalmos	1	1.9	0	0.0	^{FE}P=1.0
Distichiasis	0	0.0	2	4.3	^{FE}P=0.214
Dermolipoma	6	11.3	0	0.0	**^{FE}P=0.029***
Telecanthus	2	3.8	0	0.0	^{FE}P=0.497

X^2: Chi-Square test ^{FE}P: Fisher's Exact test *significant at P≤0.05

Regarding laterality, the ninety nine eyes with congenital and developmental eyelid anomalies showed that a total of 45 eyes were right (45.5%) and 54 eyes were left (54.5%). There was a significant prevalence of colobomas in left eyes and dermolipoma in right eyes. (Table 10)

Table (10): Showing laterality among eyes with detected eyelid anomalies.

Eyelid anomalies	Studied eyes (n=99)				Significance
	Right (n=45)		Left (n=54)		
	No.	%	No.	%	
Ptosis	21	46.7	33	61.1	X^2=2.07 P=0.151
Colobomas	2	4.4	10	18.5	**X^2=4.56 P=0.033***
Int.&Ext. A.Dermoid	10	22.2	5	9.3	X^2=3.21 P=0.073
Hemangioma	0	0.0	1	2.2	^{FE}P=1.0
Symblepharon	1	2.2	1	1.9	^{FE}P=1.0
Kohn Romano	2	4.4	2	3.7	^{FE}P=1.0
Lagophthalmos	1	1.9	0	0.0	^{FE}P=1.0
Distichiasis	1	2.2	1	1.9	^{FE}P=1.0
Dermolipoma	6	13.3	0	0.0	**^{FE}P=0.007***
Telecanthus	1	2.2	1	1.9	^{FE}P=1.0

X^2: Chi-Square test ^{FE}P: Fisher's Exact test *significant at P≤0.05

Specific meta-analysis for ptosis in affected eyes (n=54)

Among the examined 54 ptotic eyes, visual acuity was found to range from (-1.25) to (+4) which has a mean of 2.3 and SD ±2.2. Palperal fissure ranged from 2-10mm with a mean of 5.1 and SD ±1.9. Eyelid crease ranged from 0-7mm with a mean of 2.4 and SD ±2.5. Marginal reflex distance ranged from 0-4mm with a mean of 1.3 and SD ±0.9. Levator function fluctuated between 0-12mm with a mean of 5.5 and SD ±3.1 respectively. (Table11)

Table (11): Showing clinical characteristics of the studied ptotic eyes. (n=54)

Clinical characteristics	Studied eyes (n=54)		
	Min-Max	Mean±SD	Median (Q1-Q3)
Refraction [n=52]	-1.25 – 4	2.3±2.2	2 (0.7-3.2)
Palberal fissure (mm) [n=54]	2-10	5.1±1.9	5 (4-6)
Eye lid crease (mm) [n=54]	0-7	2.4±2.5	3 (0-4)
MRD (mm) [n=54]	0-4	1.3±0.9	2 (0-2)
Levator action (mm)[n=52]	0-12	5.5±3.1	5 (3-8)

Among the studied ptotic eyes, 2 (3.7%) were found to have jaw winking while 52 (96.3%) were found free. (Table 12)

Table (12): Showing presence of jaw winking in the studied ptotic eyes.

Clinical characteristics	Studied eyes (n=54)	
	No.	%
Jaw winking [n=54]		
Present	2	3.7
Absent	52	96.3

Treatment prospective of 80 affected cases (n=80)

This study was done on 9484 cases, of which 80 cases were found to have an eyelid anomaly. Fifty eight cases (72.5%) of affected cases didn't need any surgical intervention at that time and were directed to follow up. Surgical and medical interventions were done for 22 cases (27.5%) of affected cases.

Thankfully, the success rate of the surgical intervention is 22 cases (100%) with the patients being followed up for 3-6 months post operatively. (Table 13)

Table (13): Case management scale.

Examined	Diseased	Follow up	Treatment	Success rate
9484	80	58	22	22

Figure (12): Right upper eyelid coloboma.

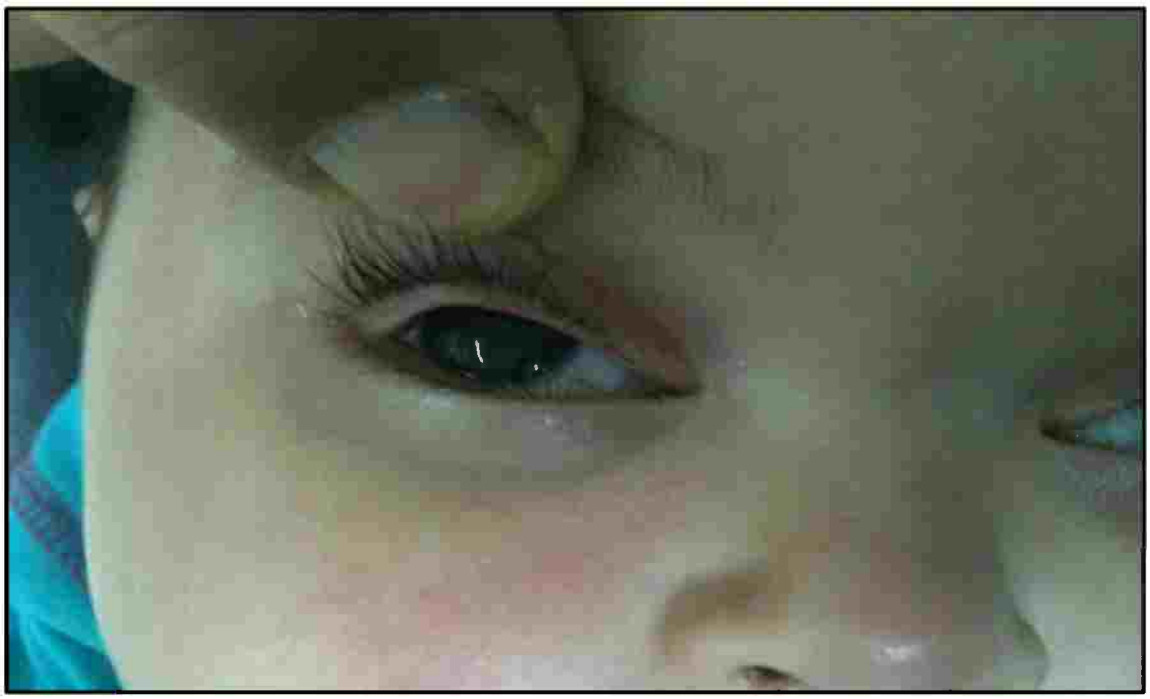

Figure (13): Right lower lid Districhiasis.

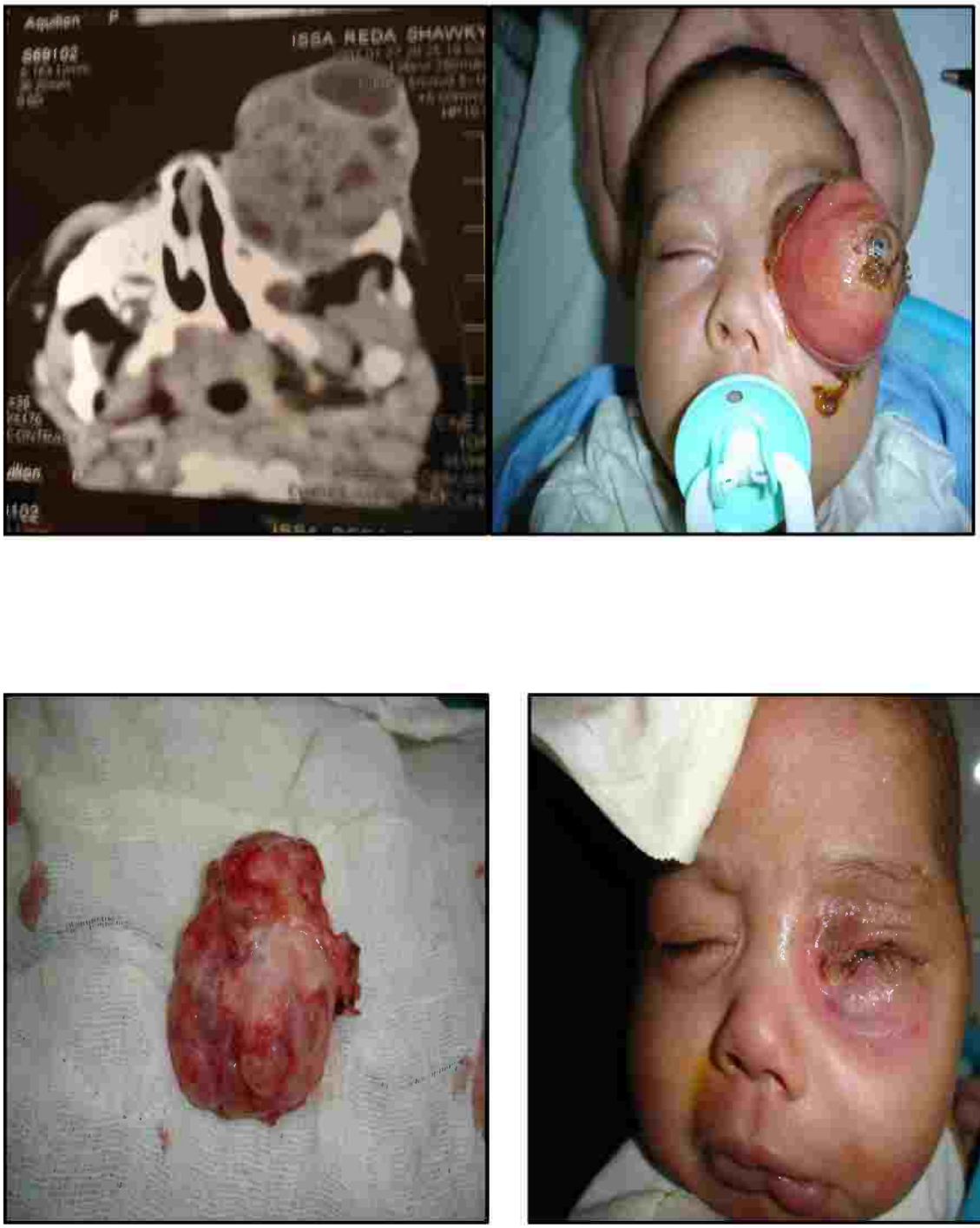

Figure (14): Teratoma tumor diagnosed and excised with post-operative picture.

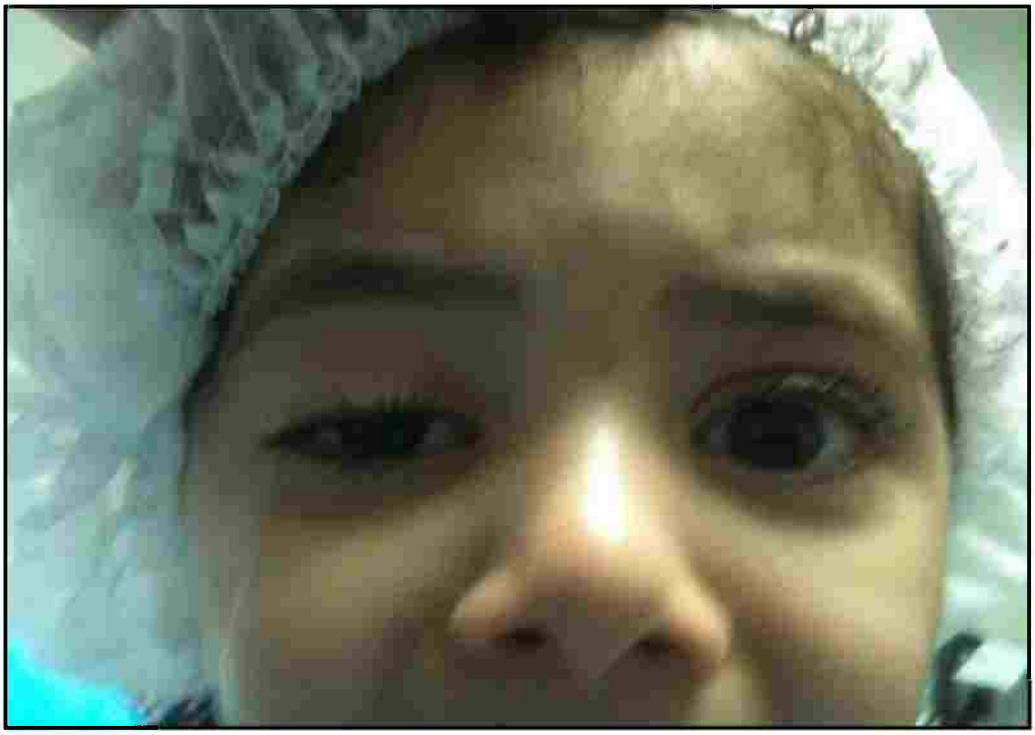

Figure (15): Upper eyelid ptosis in right eye before correction.

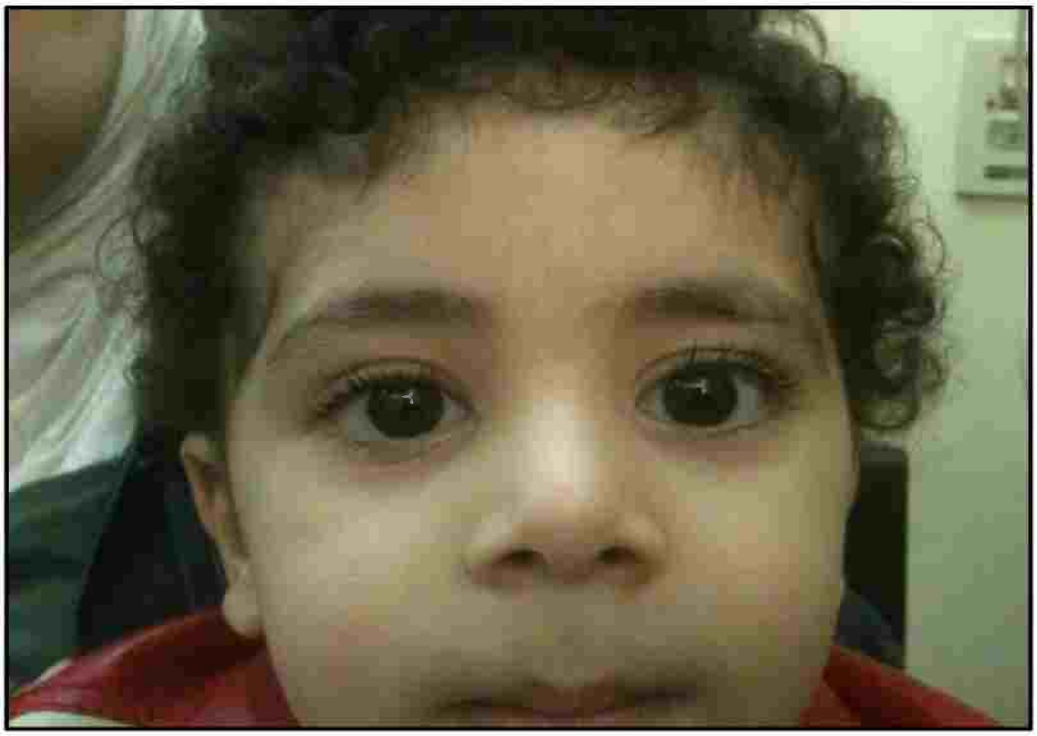

Figure (16): Right upper eyelid post ptosis correction surgery.

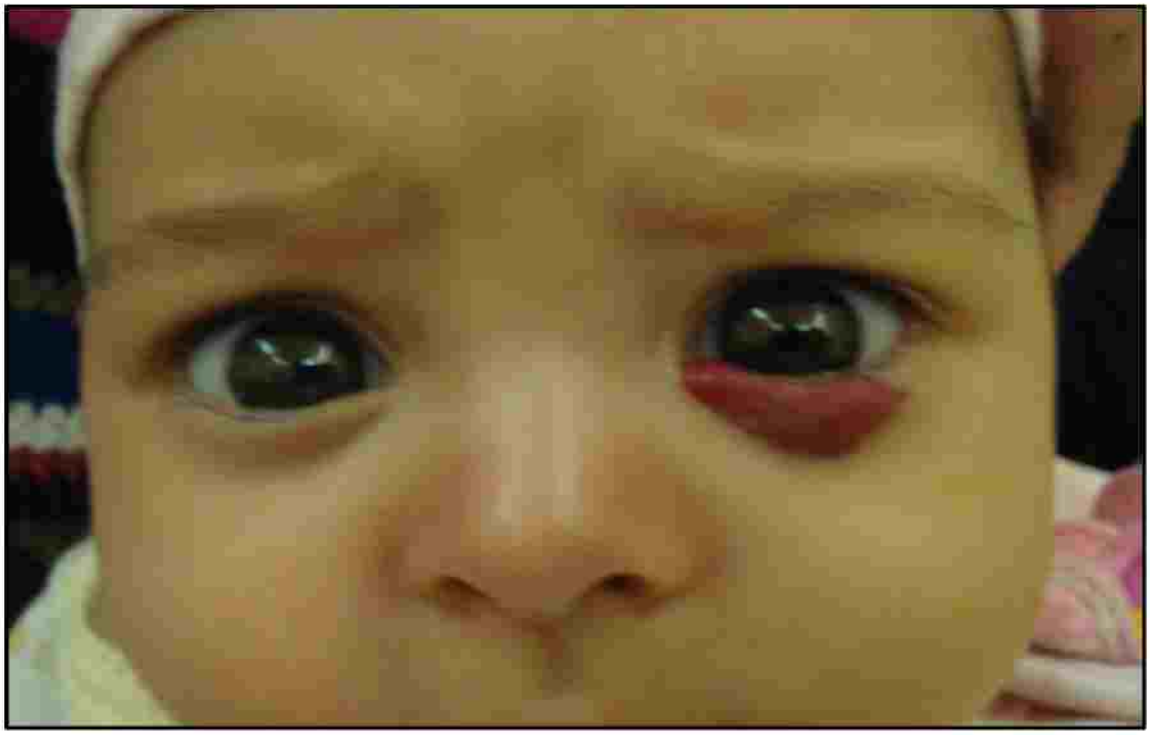

Figure (17): Lower eyelid hemangiomas in left eye.

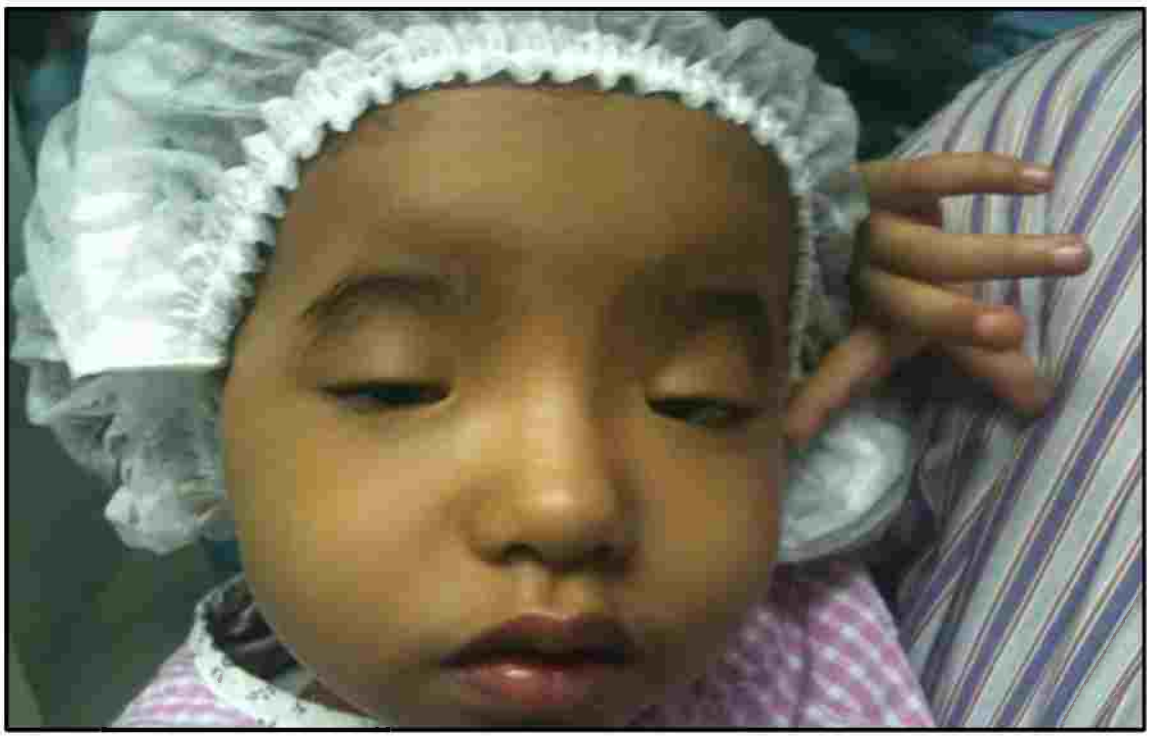

Figure (18): Blepharophimosis-Ptosis-Epicanthus Inversus (BPES).

Figure (18): A case of hemangioma in left eye treated by oral propranolol (Courtesy Prof Dr. Hisham Farouk Idriss).