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- CROSS-LINKING RESULTS IN PAEDIATRIC AGE GROUP
- OCULAR HAEMANGIOBLASTOMA AND RENAL CELL CARCINOMA IN VON HIPPEL-LINDAU DISEASE

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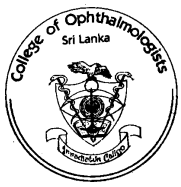

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Journal Articles

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2. Smith JD, Jones TS. Ophthalmology and society. *Surv Ophthalmol* 1997; **42**: 65-78.

Books

3. Smith JD, Jones TS. Public JQ, et al. Ophthalmology and the World. Boston. Bayside Press, 1997, pp 1-9.

Chapters

4. Stevens JT. A transcendentalist's view of optics, in Smith JD (ed): Ophthalmology and the Universe, Vol. 6. Part 3. Boston, Bayside Press, 1997. ed 2, pp 230-245.

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VOL. 22 | 2016 | NO. 2

CONTENTS

Two-year corneal cross-linking results in paediatric age group with documented progressive keratoconus

I. K. Devasurendra[1], R. S. Walpitagamage[3], D. M. M. A. K. Dissanayake[3], D. M. S. V. Dissanayake[2], K. Jeykanth[2], D. H. H. Wariyapola[4]

The Journal of the College of Ophthalmologists of Sri Lanka 2016; 22: 65-68

Abstract

Introduction and Objectives: *Keratoconus is most frequently diagnosed after adolescence, but the corneal ectatic process starts at a much younger age. Corneal CXL with riboflavin and UVA is a technique of corneal tissue strengthening using riboflavin as a photosensitizer and UVA to increase the formation of intra- and inter-fibrillar covalent bonds by photosensitized oxidation thus stabilizing or slowing down the progression. Our main objective was to assess uncorrected visual acuity (UCVA), best corrected visual acuity (BCVA), cylindrical power and Keratometer readings (Kmax and Kmin) at 1 month, 3 months, 6 months, 1 year and 2 yrs in relation to pre op readings. In addition to that we have compared the mean parameters between the total and the sub-group of under 15 years.*

Methodology: *Patients with progressive moderate to severe keratoconus. (K max 59.5-47.9) were analyzed retrospectively in this study. The UCVA, BCVA, Refraction, K readings and topography were analyzed in 16 eyes of 12 patients within the age group of 11 to 17 years with confirmed keratoconus who underwent CXL during the period of 2014-2016 at Nawaloka Hospital. Patients who completed a minimum of 1 month follow-up after the procedure were included (Mean follow up 17.93 months). The procedure was performed by a single Consultant Eye Surgeon.*

Results and Conclusion: *Out of the 12 patients the M:F ratio was 3:1. The age ranged from 11 to 17 years with the mean age being 14.1875 yrs (Table 1).62.5% of the age group had bilateral disease. Pre procedure mean K max and K min were 53.1188 and 47.6594 respectively and the mean cylinder was 3.4038. Post procedure K values were 53.57 and 46.01 and the cylinder was 3.25 at 2 years thus showing a significant improvement. At mean follow up of 17.93 months after crosslinking, there was a reduction of mean keratometry values by 1.65 D in the flattest meridian and a minimal steepening of 0.45D in the steepest meridian. This was associated with an improvement in visual acuity and the amount of astigmatism. Higher rate of progression observed in less than 15 yrs age group compared to total population but this could also be slowed by CXL.*

Introduction

The first documented study on collagen cross linking was a pilot study by Wollensak *et al*. (1), on humans (23 eyes of 22 keratoconic patients) started in 1998 and results demonstrating successful halting of keratoconus progression were published in 2003. After that there were several studies on this topic. Recently CXL has been used as an intervention in pediatric keratoconus.

Keratoconus is most frequently diagnosed after adolescence, but the corneal ectasia process starts at a much younger age. Studies have shown that pediatric keratoconus demonstrates a higher rate and speed of progression as compared to adult keratoconus. Léoni-Mesplié *et al*. (2), conducted a retrospective study to assess the severity of keratoconus at diagnosis and its progression over a period of 2 years in children compared to adults. Keratoconus in children was significantly more severe at diagnosis, with 27.8% being stage 4 vs 7.8% of adults and keratoconus evolved faster in children as compared with adult group. In addition children with keratoconus are frequent eye rubbers, especially the subgroup of children with coexisting vernal keratoconjunctivitis (VKC). Therefore keratoconus progression in children is aggressive and may not halt on its own. This may lead to progressive visual impairment in pediatric patients endangering their social as well as educational development, thus affecting their quality of life.

Early treatment could stabilize the disease or prevent advanced disease thus preventing or delaying potential corneal transplantation, as corneal transplantation in children carries a poorer prognosis than in adults(3,4). CXL is effective in halting the progression of keratoconus with an excellent safety profile in adults. For this reason, CXL has been recently utilized and evaluated in children.

Objectives and Methodology

Objectives 1) To assess UCVA, BCVA, refraction and K readings (Kmax and Kmin) at 1 month, 3 months, 6 months, 1 year and 2 yrs in relation to pre op readings.

[1]*Senior Registrar in Ophthalmology,* [2]*Registrar in Ophthalmology,* [3]*Medical Officer,* [4]*Consultant Eye Surgeon, Sri Jayawardanepura General Hospital, Kotte, Sri Lanka.*

2) To compare the mean parameters between the total and the sub-group of under 15 years.

Method: Retrospective case series analysis. Patients with progressive moderate to severe keratoconus. (K max 59.5-47.9) were taken into the study.

The UCVA, BCVA, refraction, K readings and topography were analyzed in 16 eyes of 12 patients within the age group of 11 to 17 years with confirmed keratoconus who underwent CXL during the period of 2014-2016 done at Nawaloka Hospital. Patients who completed a minimum of 1 month follow-up after the procedure were included. (Mean follow up 17.93 months) The procedure was performed by a single Consultant Eye Surgeon.

Results

Out of the 12 patients the M:F ratio was 3:1. The age ranged from 11 to 17 years with the mean age being 14.1875 yrs (Figure 1).

62.5% of the age group had bilateral disease. Pre procedure mean K max and K min were 53.1188 and 47.6594 respectively and the mean cylinder was 3.4038. Post procedure K max increased slightly during the initial six months after which it reduced marginally at 1 year. It again rose slightly but the overall rise at 2 yrs was 0.45 diopters compared to pre-op value (Figure 2). Average K min also showed a similar trend but the 2 yr mean value was 1.6 D less than the pre-op value (Figure 3). Mean cylinder came down from a pre op value of 3.4038 to a 2yr value of 3.25 D (Figure 4). In all 3 parameters the reduction was mostly observed at 1 yr.

Within the sub group of under 15 yrs K max was observed to be higher compared to the total analyzed pediatric population indicating more severe keratoconus in the former. K max progression was also higher in less than 15 yrs, but again stabilized towards 2 yrs (Figure 5). Mean K min also behaved in a similar manner. Mean cylinder fluctuated in both but showed a reduction at 1 yr compared to pre op values (Figure 7). Significant improvement in visual acuity was seen during the follow up of individual cases. Demarcation line was observed in >90% (93.78v - 15 eyes).

What happened to fellow eyes? There were 5 normal fellow eyes, 8 cross linked eyes and 2 eyes have undergone DALK. There was single borderline case followed up at clinic.

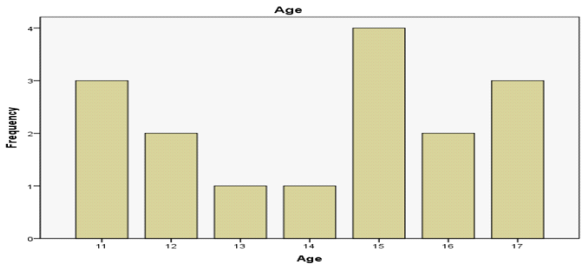
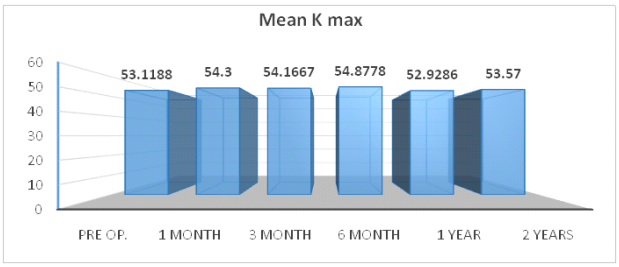


Figure 1



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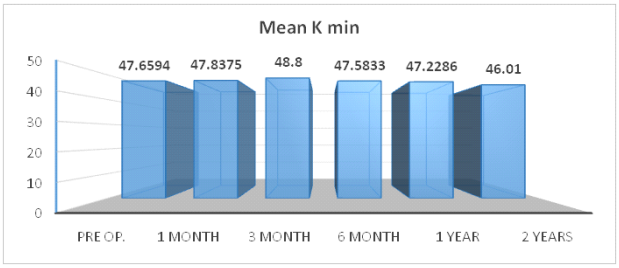

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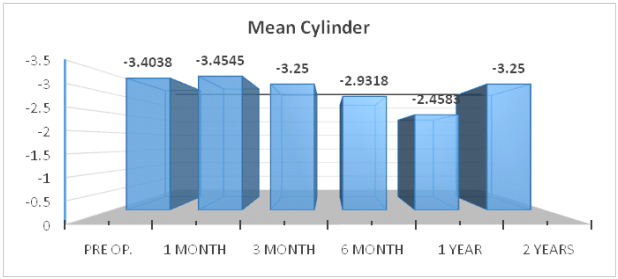

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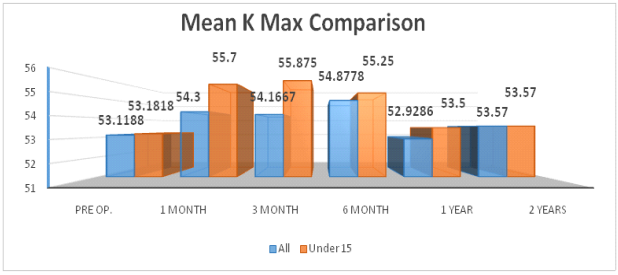

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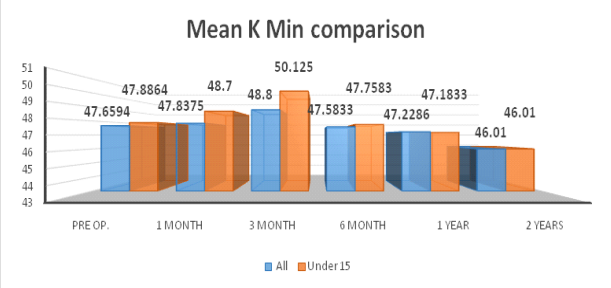

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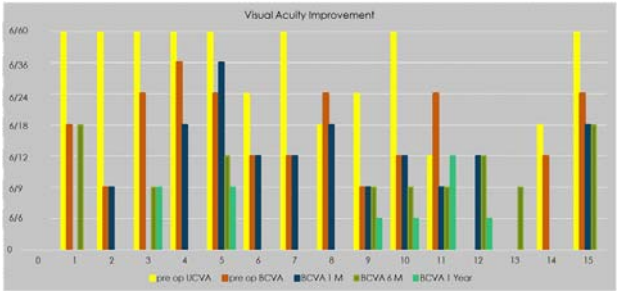

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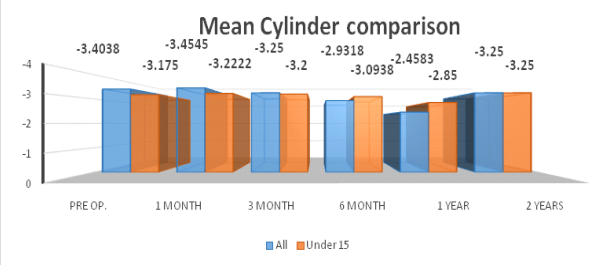


Figure 7

Demarcation line

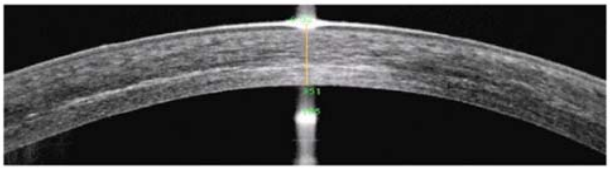
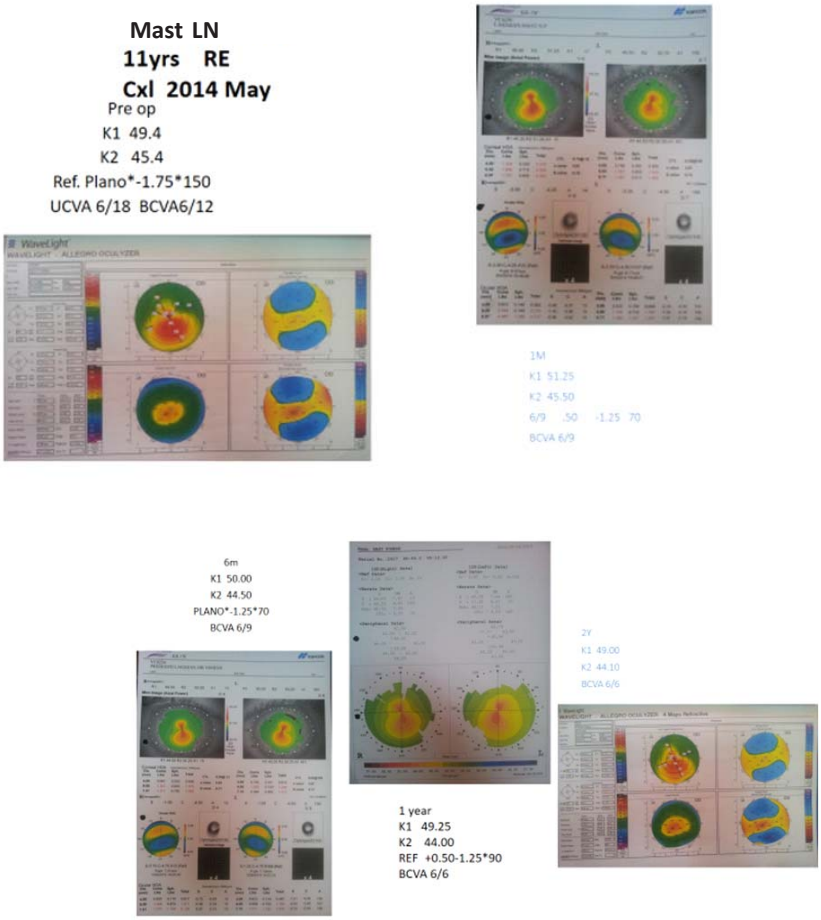


Figure 9

Case 1



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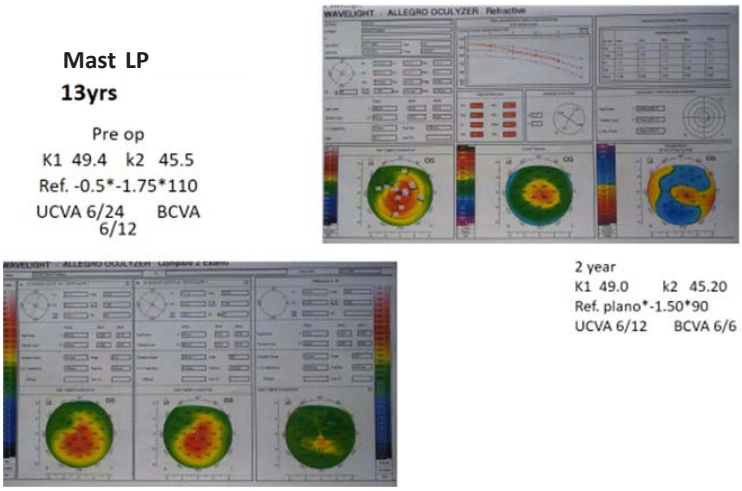

Conclusion

Major concern comprise of accelerated progression of keratoconus in pediatric age group. Collagen cross linking could stabilize or slow down the disease course and this was associated with improvement in best corrected visual acuity and the amount of astigmatism. Higher rate of progression was observed in less than 15 years, but this could also be slowed down by collagen cross linking. Thus collagen cross linking can be considered as an effective method in slowing or halting the progression of keratoconus, hence decrease the risk of amblyopia, fit of contact lens and deter an early keratoplasty.

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Traditional versus Barrett Universal II IOL calculation formula for meniscus optic IOL; a retrospective analysis

D. M. S. V. Dissanayake, D. Wariyapola, I. K. Dewasurendra, D. M. M. A. K. Dissanayake, R. S. Walpitagamage, S. K. G. S. Kurera

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Abstract

Introduction: *Despite the advances in IOL power prediction formulae, the high myopic eye has constantly been a challenge to the ophthalmologist. A multitude of factors affect the IOL power calculation in these eyes. The meniscus (concavo-convex) design of the IOL optic in +5.0D or less powers, is an important, yet disdained, factor which affects the effective lens position. The Barrett Universal II IOL calculation formula is one that is specifically designed for meniscus design IOLs.*

This paper presents retrospective single-center, single-surgeon data of high myopic patients implanted with meniscus lenses. Post-operative refraction at 4 weeks was compared among the predicted refractive error for the implanted IOL for SRK-T, Holladay I, Hoffer-Q, Haigis and Barrett Universal II formulae.

Results: *When calculated with the manufacturer provided IOL constants, the mean deviation of the error predicted by the formula from post-operative value was 1.13D for Barrett Universal II formula, 1.85D for SRK/T, 2.19D for Holladay 1, 2.44D for Hoffer Q and 1.66D for Haigis formula. But when the ULIB optimized IOL constants were used for the traditional formulae, deviations from the post-operative values were 1.02D, 1.00D, 1.00D and 1.10D for the SRK/T, Holladay I, Hoffer Q and Haigis respectively.*

Conclusion: *The Barrett Universal II formula is highly accurate for high myopic eyes. Equivalent accuracy can be achieved if ULIB optimized values are used for the traditional formulae.*

Introduction

The prediction of intraocular lens (IOL) power for cataract surgery is a task the ophthalmologist has begun to master since of recent. Increase in the accuracy of biometric data, such as with the use of laser interference biometry, increases in the biometric parameters used in the calculation, consideration of surgeon factor and more advanced IOL calculation formulae have aided in this endeavor.

Successive generations of IOL calculation formulae have demonstrated increasing accuracy of power prediction, and, improve the range of axial lengths for which the formula can be successfully applied. However, eyes that deviate significantly from the normal population, such as the high myopes and high hypermetropes, still have less accuracy with the commonly used IOL formulae.

The commonly used IOL formulae are the Hoffer Q, Holladay I, SRK/T and Haigis. Their accuracy differs with the change in axial length, and so the formula of choice varies according to the axial length (Table 1) (2).

Table 1. formula of choice according to the axial length of the eye

Axial length	*Formula of choice*
< 20 mm	Hoffer Q / Holladay II
20-22 mm	Hoffer Q
22-24.5 mm	SRK/T, Hoffer Q, Holladay
24.5-26.0 mm	Holladay I
> 26.0 mm	SRK/T

In addition to the error of the IOL formula, the high myopic eyes are hampered by a second variable that affects the post-operative vision. Very low power intraocular lenses (+5.0D or less) employ a meniscus design of the optic, while others have a biconvex design. The principle planes and the nodal points of this optic design are positioned differently, when compared to that of a biconvex lens of the same power. Thus, meniscus design IOLs have different optical properties (Figure 1), a factor that needs to be considered when calculating for IOL power. However the commonly used 3rd generation IOL formulae do not consider the effect of the optic design in their calculation.

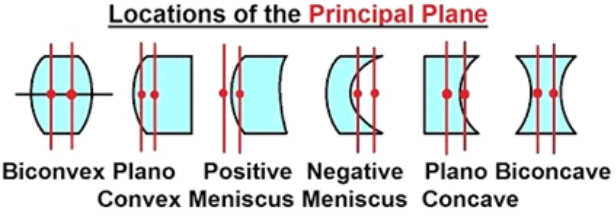

Figure 1. *Lens designs and their principal planes.*

The Barrett Universal II formula is a 3rd generation IOL power calculation formula that considers the optic design of the IOL to be implanted. This formula promises improved post-operative refractive outcomes due to this reason[1].

Pre-operative and post-operative data gathered from patients undergoing phacoemulsification in selected centers from all over the globe is collected through the ULIB database (User group for Laser Interference Biometry) (3). This database compiles data obtained through optical biometry using laser interferometry. One of ULIB's special concerns is the optimization of lens constants for the calculation of IOL power.

The ULIB recommends alternative A-constant values for the common IOL calculation formulae based on its data. For the Alcon MA60MA lens, the recommended values when measured via IOLMaster biometry are as follows (Table 2).

Objective

To compare the accuracy of the Hoffer Q, Holladay I, SRK/T and Haigis formulae using the manufacturer provided IOL parameters and with ULIB optimized parameters, against Barrett Universal II formula using manufacturer provided IOL parameters.

Methodology

Retrospective analysis of 7 eyes of 6 patients was performed with the use of previous medical records. All patients were from Sri Jayewardenapura General Hospital and operated by a single surgeon, Dr. D.H.H. Wariyapola. Only uncomplicated phacoemulsification surgery patients were selected.

Patients from September 1st 2015 to August 31st 2016 were studied. Eyes with axial lengths 262.00 mm or more were selected for analysis.

Patients requiring ultrasonic biometry were excluded, and all patients underwent IOLMaster 700 (Zeiss) biometry.

Pre-operative refraction and post-operative subjective refraction done at 4 weeks was used to assess the refractive error.

All selected patients were implanted with Alcon MA60MA (Expand Series) IOLs. Implanted lens power was selected using SRK/T formula with up-to-date ULIB IOL parameters. Target post-operative refractive error was kept at -1.0D except for 1 eye which required a -8.0D lens, which was not available for purchase at the time of surgery.

Using the pre-operative IOLMaster 700 optical biometry values, recalculation of IOL power was performed using Hoffer Q, Holladay I, SRK/T and Haigis formulae. The first values were obtained for the manufacturer provided lens parameters (A-constant 118.9, SF 1.73, a0 1.839263, a1 0.4, a2 0.1, pACD 5.49). The second set of values was obtained with the same formulae using the ULIB optimized lens parameters (Table 2). Calculations were performed using the free online IOL calculator (http://www.eyecalcs.com/WEBCALCS/IOLcalc/IOL.html).

The same biometry was used to calculate the lens power using the Barrett Universal II formula. IOL parameters provided by the manufacturer were used. The free online calculator was used for calculations (http://www.apacrs.org/barrett_universal2).

In each step, the predicted post-operative error for the already implanted IOL was obtained. This was then compared to the post-operative subjective refraction value. Spherical equivalence of the post-operative refraction was used for comparison.

Results

Of the 6 patients 2 were males. Age ranged from 51 years to 64 years.

Table 2. Comparison of parameters provided by the manufacturer and the ULIB optimized values

Parameters provided by the manufacturer	*Lens*	*ULIB recommended value for IOL Master*			
		Haigis	*Hoffer Q*	*Holladay 1*	*SRK/T*
A constant = 118.9 Sf = 1.73 pACD = 5.49 a0 = 1.839263 a1 = 0.4 a2 = 0.1	MA60MA +diopter lens	a0 = 5.78 a1 = 0.40 a2 = 0.10	pACD = 15.94	sf = 10.29	A = 126.6
	MA60MA -diopter lens	a0 = -4.22 a1 = 0.40 a2 = 0.10	pACD = -5.52	sf = -7.08	A = 103.6

The axial lengths ranged from 26.58 to 36.58 and the K readings ranged from 42.24 to 45.12. Anterior chamber depth ranged from 3.1 mm to 4.13 mm.

The lens powers implanted ranged from +5.0 to -4.0D with only two eyes requiring negative powers (-4.0D and -5.0D). One patient required a -8.0D lens but was implanted with a -5.0D lens due to lack of availability. Post-operative refractive error ranged from +1.35 to -1.5D in spherical equivalence. The best corrected visual acuity ranged from 6/9 to 6/36.

With the manufacturer provided IOL parameters for the power calculation formulae, the expected refractive error and the post-operative refractive error is as given in the Table 3.

The deviation of the error predicted by formulae from the post-operative value was obtained for each instance. The absolute value of deviation was considered in the calculation. This is given in Table 4. If relative deviation was used a positive value in one eye will cancel negative value in another eye and falsely decrease the total deviation value.

When the ULIB optimized IOL parameters (Table 2) were used for the calculation with the traditional formulae, the predicted error for the implanted IOL was as given in Table 5.

The deviation of this predicted value was calculated, as previously done. The absolute deviation was obtained. These are given in Table 6

Table 3. The predicted post-operative refractive error when using manufacturer parameters for calculation

Eye	*Implanted IOL power (Diopters) refraction*	*Spherical equivalence of post-op*	*Error for Barrett Universal II*	*Error for SRK/T*	*Error for Holladay 1*	*Error for Hoffer Q*	*Error for Haigis*
1	- 4.0	-1.50	-2.11	-3.6	-3.68	-4.21	-3.23
2	+ 2.0	-1.00	-0.72	-1.29	-1.7	-1.86	-1.12
3	+ 2.0	-0.25	-1.21	-1.77	-2.2	-2.38	-1.67
4	+ 4.0	+0.875	-0.67	-0.8	-1.25	-1.28	-0.58
5	+ 2.0	-0.50	-0.69	-1.31	-1.72	-1.91	-1.2
6	- 5.0	+1.375	-2.66	-4.35	-4.38	-4.98	-4
7	+ 4.0	-1.00	-0.28	-0.84	-1.37	-1.44	-0.8

Table 4. The deviation of the value predicted by each formula from the actual value

Eye	*IOL used*	*Spherical equivalence of post-op refraction*	*Error for Barrett Universal II*	*Error for SRK/T*	*Error for Holladay 1*	*Error for Hoffer Q*	*Error for Haigis*
1	- 4.0	-1.5	0.61	2.1	2.18	2.71	1.73
2	+ 2.0	-1	0.28	0.29	0.7	0.86	0.12
3	+ 2.0	-0.25	0.96	1.52	1.95	2.13	1.42
4	+ 4.0	0.875	1.545	1.675	2.125	2.155	1.455
5	+ 2.0	-0.5	0.19	0.81	1.22	1.41	0.7
6	- 5.0	1.375	4.035	5.725	5.755	6.355	5.375
7	+ 4.0	-1	0.28	0.84	1.37	1.44	0.8
Sum of deviation			7.9	12.96	15.3	17.06	11.6
Average deviation			1.128571	1.851429	2.185714	2.437143	1.657143

Table 5. Predicted error for the implanted IOL when calculated with ULIB optimized parameters for the traditional formulae

Eye	*IOL used*	*Spherical equivalence of post-op refraction*	*Error for Barrett Universal II*	*Error for SRK/T*	*Error for Holladay 1*	*Error for Hoffer Q*	*Error for Haigis*
1	-4	-1.5	-2.11	-1.4	-1.6	-1.55	-1.88
2	2	-1	-0.72	-0.85	-0.95	-1.02	-0.75
3	2	-0.25	-1.21	-1.33	-1.44	-1.53	-1.28
4	4	0.875	-0.67	-0.37	-0.23	-0.1	-0.41
5	2	-0.5	-0.69	-0.87	-0.97	-1.07	-0.81
6	-5	1.375	-2.66	-1.64	-1.81	-1.7	-2.33
7	4	-1	-1.28	0.21	-0.07	0.04	-0.26

Table 6. The deviation of the predicted error from the post-operative spherical equivalence, when the traditional formulae were applied with ULIB optimized IOL parameters

Eye	*IOL used*	*Spherical equivalence of post-op refraction*	*Error for Barrett Universal II*	*Error for SRK/T*	*Error for Holladay 1*	*Error for Hoffer Q*	*Error for Haigis*
1	-4	-1.5	-0.61	0.1	-0.1	-0.05	-0.38
2	2	-1	0.28	0.15	0.05	-0.02	0.25
3	2	-0.25	-0.96	-1.08	-1.19	-1.28	-1.03
4	4	0.875	-1.545	-1.245	-1.105	-0.975	-1.285
5	2	-0.5	-0.19	-0.37	-0.47	-0.57	-0.31
6	-5	1.375	-4.035	-3.015	-3.185	-3.075	-3.705
7	4	-1	-0.28	0.1	-0.1	-0.05	-0.38
Sum of deviation		7.9	7.17	7.03	7.01	7.7	
Average deviation		1.128571	1.024286	1.004286	1.001429	1.1	

Discussion

On analysis of the deviation of the predicted refractive error from the post-operative value (Table 2), the average deviation from post-operative value was 1.13D for Barrett Universal II formula, 1.85D for SRK/T, 2.19D for Holladay 1, 2.44D for Hoffer Q and 1.66D for Haigis formula.

All 7 eyes showed higher accuracy for Barrett Universal II formula, when manufacturer provided IOL constants were employed for the calculation (Figure 2). Of the traditional 3rd generation formulae, the SRK/T and the Haigis were more accurate than the Holladay I and Hoffer Q.

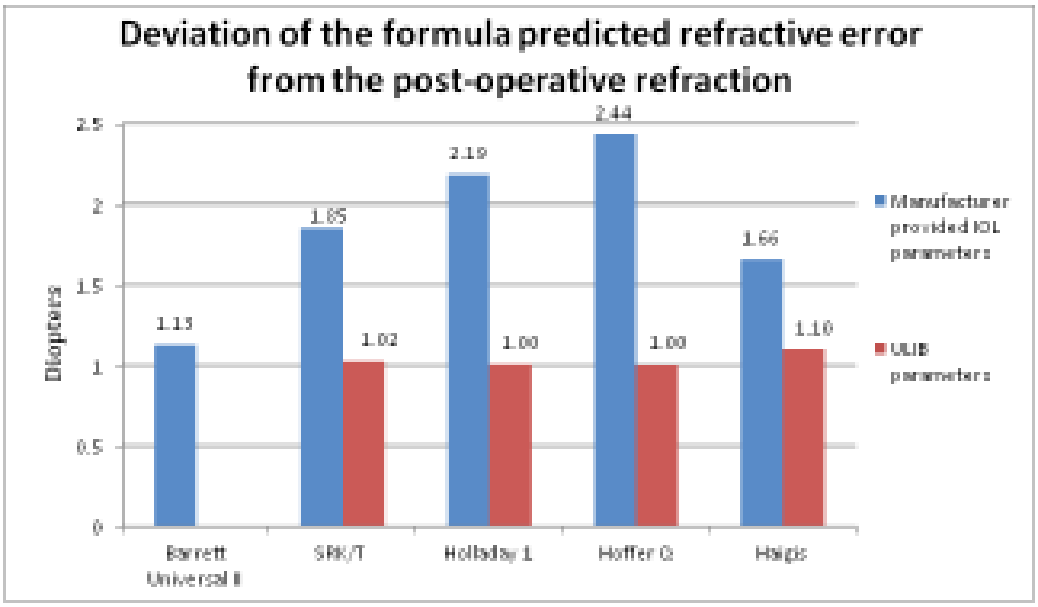

Figure 2. *Comparison of the deviations from the post-operative subjective refraction when calculated with manufacturer derived IOL parameters and ULIB optimized parameters.*

But when the ULIB optimized IOL parameters were utilized for the calculation of the IOL power, the traditional formulae were superior to the Barrett Universal II formula (Table 4). The deviations from the post-operative values were 1.02D, 1.00D, 1.00D and 1.10D for the SRK/T, Holladay I, Hoffer Q and Haigis formulae respectively. All of these values were lower than the Barrett Universal II value of 1.13D. (Figure 2)

The above average deviation values show about 1 diopter difference from the post-op subjective refraction. This value would have been much lower if eye number 6 was not considered. The eye number 6 is the largest eye with an axial length of 36.58 mm. Without the consideration of this eye the results of all the formulae are very similar with the Barrett Universal II having 0.644D error, SRK/T 0.693, Holladay 1 0.641, Hoffer Q 0.656 and Haigis having a 0.666 diopter errors. The patient with eye number 6 was implanted with a -5.0 Diopter IOL due to the lack of availability of other IOL powers at the time of surgery. This eye was over corrected and required a plus diopter lens correction post-operatively.

This study has several limitations. The sample size is small as only 7 eyes were found to fall within the study criteria. This precludes statistical analysis.

The study does not involve thin biconvex lenses, and only eyes implanted with meniscus lenses were involved in the study. These lenses have limited availability in Sri Lanka and they are not available in half diopter increments.

Only the Alcon MA60MA lens was considered in this study. The results may differ for other lens brands.

Conclusion

The Barrett Universal II formula shows higher levels of accuracy for the high myopic eyes, when the IOL constants provided by the manufacturer are used for the calculation. These results are compatible with the published literature (4,5).

However when ULIB optimized IOL constants are used the traditional formulae also show equivalent or better accuracy. If the optical biometry axial length value was further optimized, the accuracy may have been superior to that found in this study (6).

The Barrett Universal II formula is easy to use and is freely available online. Currently it doesn't have ULIB optimized values. This formula does not require axial length optimization of the optical biometry reading.

It is important that the surgeon go through the A-constant and other IOL parameters used for power calculation as well as the predicted IOL powers, when reviewing the biometry data sheet pre-operatively. This is of special importance in the high myopic eye. Having up-to-date ULIB optimized lens constants installed in the optical biometry machines and using the most appropriate formula will allow the surgeon to obtain the most accurate lens power calculation, and so, the best post-operative refractive outcome, in the high myopic eye.

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Comparision of spectral domain anterior segment optical coherence tomography and ultrasound biomicroscopy in assesment of anterior chamber depth and anterior chamber angle

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Introduction

Primary angle closure glaucoma has a rising prevalence in ageing population (1). The risk of getting primary angle closure is dependent on anatomical features of the anterior segment. The anterior chamber depth, position of ciliary processes and iris thickness are associated (2). These features are difficult to distinguish in gonioscopy and hence require better methods of evaluation (3) (4).

The two main methods that are currently being used are the anterior segment spectral domain optical coherence tomography (AS OCT) and the ultrasound biomicroscopy(UBM). The angle open distance is measured in both methods at 500 µm (AOD 500), at 750 µm(AOD 750) as well as the trabecular iris angle (TIA) (5). Ultrasound biomicroscopy has the further advantage of detecting anatomical and pathological defects posterior to the iris, which can lead to secondary angle closure.

We have compared the results of anterior segment OCT and ultrasound biometry in ten eyes with suspected narrow angles and in ten eyes with clinically normal angled eyes, in order to compare the two methods of angle evaluation.

Method

Twenty eyes were used in our study in two groups: one group who had an open angle during clinical evaluation with slit lamp and gonioscopic examination and the other group with a narrow angle (grade 1 and 2 with van Herick method and grade 1 and 2 with the Scheie method). Clinical grading was performed by a single examiner. 10 eyes with clinically open angles and 10 eyes with clinically narrow angles with no evidence of glaucoma were recruited and evaluated separately with the AS OCT and UBM. The following four measurements were obtained.

1. AOD 500 as the distance from iris to cornea at 500µm from scleral spur. Measurements were obtained in the superior, inferior, nasal and temporal quadrants separately.
2. ACD-anterior chamber depth.

Results

The mean values of AOD 500 measurements in both clinically open angled and narrow angled patient groups with AS OCT and UBM summarized below.

Table 1.1. The mean value of AOD 500 in the four quadrants of clinically open angled patients and clinically narrow angled patients measured with ASOCT and UBM

	Clinically narrow angled		*Clinically open angled*	
	AS OCT	UBM	AS OCT	UBM
Inferior	0.145	0.165	0.289	0.341
Superior	0.124	0.125	0.276	0.282
Temporal	0.151	0.156	0.289	0.305
Nasal	0.140	0.155	0.276	0.282

The mean value of the ACD measurements are as follows for the two groups of patients using AS OCT and UBM.

Table 1.2. The mean values of ACD of clinically open angled and clinically narrow angled patients using AS OCT and UBM

	Clinically narrow angled		*Clinically open angled*	
	AS OCT	UBM	AS OCT	UBM
Mean ACD	1.972mm	1.827mm	2.708mm	2.316mm

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Discussion

In our study we were able to identify correctly patients with narrow angles and open angles with both the UBM and the AS OCT (Table 1.1.). Previous studies have also identified that both methods are similar in screening and follow up of primary angle closure glaucoma using measurements such as AOD 750 (4). The clinically narrow angled group had OD500 values within the reference range of previous studies (2).

However, the four quadrant values obtained by the UBM and AS OCT show different measurements (Table 1.1.). In the narrow angle group, UBM showed the highest mean value at the inferior angle and a lowest mean value at the superior angle, while the AS OCT shows the highest mean value at the temporal angle and the lowest in the superior similar to UBM.

In the clinically open angled group, the superior and the temporal angles showed the highest mean AOD 500 value with AS OCT and the inferior angle, with UBM. The lowest mean values were in the superior and nasal quadrants when measured with the AS OCT as well as the UBM. However this is dissimilar to the values obtained in the narrow angled group.

The anterior chamber depth measurements from the two methods show that both methods were able to detect the depth in relation to the clinical finding (Table 1.2.). However the mean value were lower with the UBM than with the AS OCT, in both the wide angled and shallow angled groups. This was assumed to be caused by compression of the cornea during measurements with the UBM which is a contact based measure. Literatures not available correlate these findings.

Conclusion

Both SD-AS OCT and UBM are capable of correctly identifying narrow angles and shallow anterior chambers. However UBM has an advantage of displaying the angle recess area and structures behind the iris is important to rule out secondary causes of angle closure such as plateau iris as 21 and iridocilliary cysts.UBM seem to underestimate the depth of the anterior chamber as it compresses the eye during measurement. The best method to measure the anterior chamber depth is the non-contact method of AS OCT.

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Response of intravitreal Aflibercept – a case of Wet age related macular degeneration (Wet ARMD)

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Introduction

Choroidal neovascularization (CNV) in age related macular degeneration (ARMD) is thought to be induced by several events, such as accumulation of lipid metabolic byproducts, oxidative stress, reduction in choriocapillaris blood flow, and alterations in Bruch's membrane.

Hypoxia has been shown to induce Vascular Endothelial Growth Factor (VEGF) gene transcription. As a response to metabolic distress, the retinal pigment epithelium (RPE) and retinal tissue produce various factors, particularly VEGF, which induce Chroidal Neovascularization (CNV) proliferation.

Ranibizumab and Aflibercept, were designed specifically for the treatment of ARMD. A third drug, Bevacizumab was originally developed to treat various types of cancer, but is commonly used in patients with ARMD.

Bevacizumab is a nonspecific VEGF with 2 binding site per molecule, prevents all isoforms of VEGF-A binding to endothelial receptors.

Ranibizumab is a monoclonal antibody fragment (fab) binds to VEGF-A similar in action as Bevacizumab

Aflibercept is a recombinant fusion protein. Inhibits VEGF-A, VEGF-B and placental growth factor (PlGF). It binds to VEGF-A higher affinity than its natural receptors and acts as VEGF trap.

Case details and results

65 years old male patient with wet ARMD of his left eye has been treated with multiple intravitreal injections of anti-VEGF since 2012.

Presenting best-corrected visual acuity in right eye is 6/60 and left eye 6/36. Right eye macula is scared. Treatment started initially with Bevacizumab, and then with Ranibizumab, due to inadequate response treatment was switched to Aflibercept.

He is presently maintaining best-corrected visual acuity of 6/24 in his left eye.

Discussion

Today, the standard of care for a patient who has been identified as having wet macular degeneration is to treat with a potent anti-VEGF agent monthly until the macula is dry on OCT. All three anti-VEGF has been studied in several trails and found to be effective.

However switching from one Anti-VEGF to another should always be considered when there is inadequate response. Aflibercept may have a better outcome due to its additional mechanism of actions than other 2 of its counter parts.

Series of OCT macula pictures from the beginning of treatment showing response to different Anti-VEGF are shown in the Figure 1 in next page.

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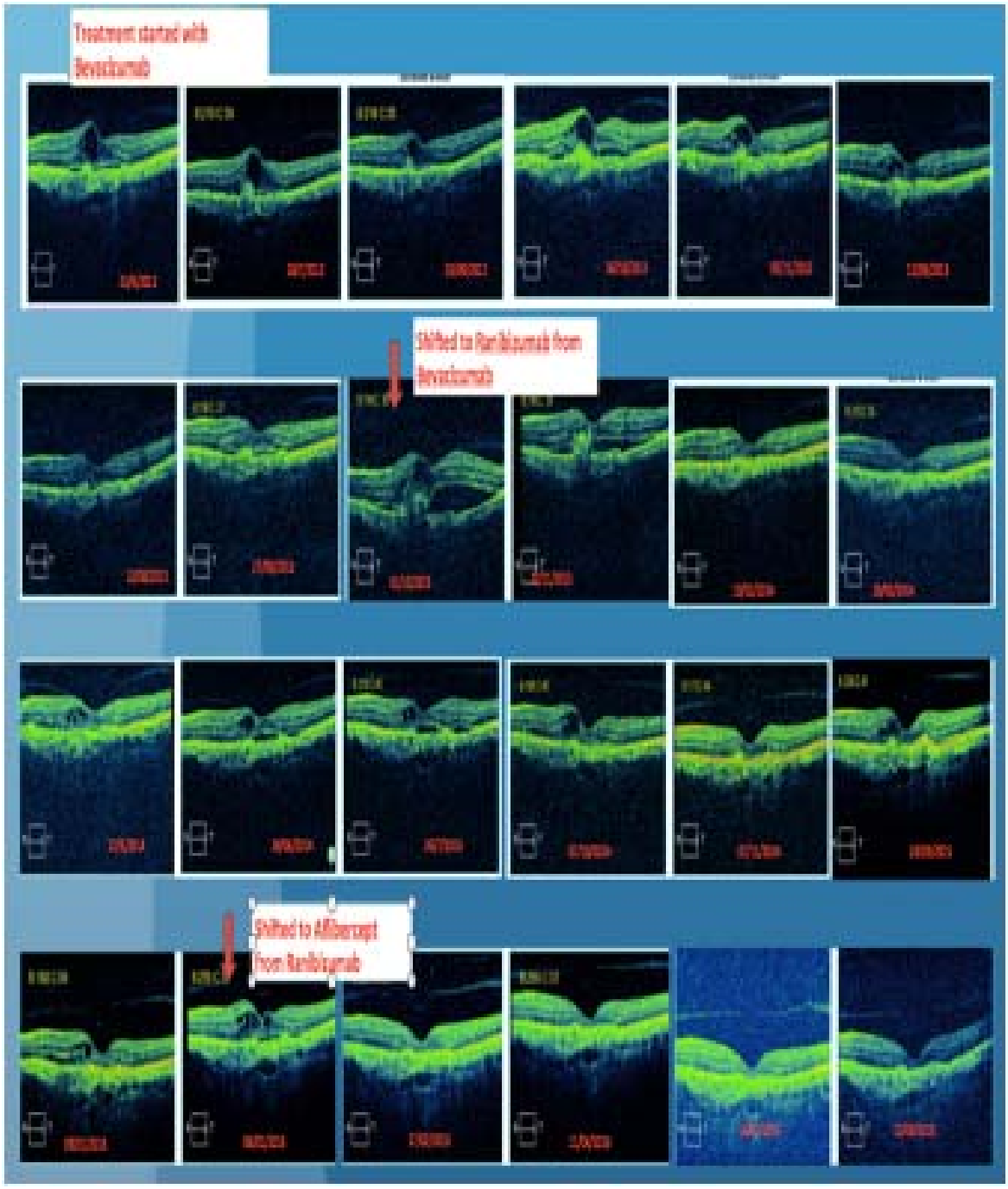
Treatment started with Bevacizumab
Shifted to Ranibizumab from Bevacizumab
Shifted to Aflibercept from Ranibizumab

Figure 1.

Improving the success of trabeculectomy

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Introduction

Traditional glaucoma surgery, such as trabeculectomy, has been performed for decades and has a proven track record of success. Unfortunately, many complications such as hypotony and slow visual recovery are known to occur and can lead to significant patient morbidity. The success of trabeculectomy depends not only on the surgery itself, but possibly even more importantly on the post-operative management.

Prevention is vastly preferable to cure, so one needs to take every precaution to avoid complications. This involves careful case selection and optimization of the operating environment prior to surgery. In addition, patients should have been counselled prior to surgery so their expectations match their post-operative experience.

Causes of failure and interventions

Scarring

The main cause of failure of trabeculectomy is the subconjunctival fibrosis. In the weeks immediately following the procedure, scar tissue forms and offers resistance to outflow. The rate at which this tissue forms depends on ethnicity, use of antimetabolites at the time of surgery and the external ocular environment (past medical therapy and previous trauma/surgery, blepharitis, and conjunctival inflammation). There is often a natural peak of resistance to outflow which then resolves with remodeling. Increases in pressure are therefore not abnormal a few weeks after a trabeculectomy.

Prevention of fibrosis greatly increases the success rates of trabeculectomy and it starts pre operatively. Mechanically reestablishing flow is the first priority. This can be achieved by ocular massage, removing releasable sutures, dividing fixed sutures (by laser) and needling of the bleb if others fail as a very last resort. Removing releasable sutures is the simplest and quickest approach by far, so give serious consideration to routine use of these sutures in your surgery.

Steroids are the next major postoperative tool to prevent scarring. Insufficient topical steroid therapy is, in my view, one of the principal causes of bleb scarring in patients who are subsequently referred to me for management.

Antimetabolites mitomycin and 5 fluorouracil are agents that inhibit fibroblasts proliferation and have been used in clinical practice. Intraoperative application of mitomycin and postoperative use of 5 FU tremendously improve the success of glaucoma filtration surgery.

Wound leak

Immediately postoperatively, all that stands between a good result and a flat anterior chamber are your flap sutures, hence the importance of care at the time of surgery. It is always better to aim to keep the IOP at around 15 to 20 mmHg on postop day one. This allows the vision to return to normal (or close to normal), so the patient can resume daily activities. In most cases pressure is slightly elevated relative to the goal, which is ideal at that point to avoid hypotony. Depending on the pressure goal, a releasable suture can be cut as early as the first day or within the first three weeks. Laser suture lysis can be performed between day 7 upto even 6 weeks or more, if intraoperative antimetabolites had been used.

Early postoperative wound leak is another major cause of failure of trabeculectomy. Clearly, if there is a button-hole at a vital site, the conjunctiva is retracting or the sutures are too loose, it needs surgical repair. I personally like suturing the conjunctiva very precisely and carefully with a mattress suture at the limbus to avoid these. There may still be a leak post-operatively during the first week or so. Studies have demonstrated such minor leaks to be of no consequence to final outcome. Larger leaks or persistent leaks are obviously more serious. There are two scenarios to consider, based on the IOP. If the leak is combined with hypotony and/or a shallow anterior chamber, then this needs careful observation and surgeons should not hesitate to re-operate to correct it. If the IOP is not too low and the anterior chamber is deep, then a more conservative approach can be considered for a period to see if matters resolve naturally.

Ocular hypotony

Early post-operative hypotony is another factor contributing to failure of trabeculectomy. It may be seen in uveitic eyes, high myopes and others with thin sclera where leakage through suture tracks is unavoidable. In such instances, it is usually a matter of 'tiding over' until the leaks seal and the ciliary body perks up with the postoperative steroids. 'Tiding over' may involve regular observation, rest (no coughing, bending, lifting

or straining), and wearing a shield at night. Should there be choroidal detachment, anterior chamber shallowing or a threat of hypotonic maculopathy, then viscoelastic or gas (SF6 or C3F8) may be used in the anterior chamber. If the hypotony is due to frank over-drainage, and especially if the anterior chamber is shallow/flat, then this is a surgical error and generally requires repeat surgical repair. It is better to face facts and do this early (on the first or second day after the operation) rather than late. Please remember that leaving an eye hypotonous increases the risk of supra choroidal-haemorrhage.

Choroidal detachments are almost always associated with hypotony, and should be evaluated from the point of prevention of hypotony. Intervention should be undertaken to prevent or resolve kissing detachments since these can be extremely destructive. Consideration of drainage or other secondary procedures are only appropriate in the extremely rare instance of exudative detachments unresponsive to medical therapy.

Infection

Infection is, fortunately, rare. Again prevention is the key. Leaks should be observed closely, protruding suture ends trimmed or removed and any surgical intervention should be covered with povidone iodine with meticulous 'no touch' technique.

Hyphaema

Hypheama is most usual on day one and settles quite rapidly. If it does not, then there is likely to be a bleeding diathesis, either pharmacological or pathological. Treat the bleeding diathesis first and await resolution.

Malignant glaucoma

In its classic form, malignant glaucoma is rare but one of the most serious complications of glaucoma filtration surgery in patients with narrow-angle or angle-closure glaucoma. This should be anticipated as a risk in short eyes (axial length <20 mm or shallow anterior chambers) and prophylactic atropine 1% should be prescribed pre-operatively and once daily for a minimum of three weeks post-operatively. Also malignant glaucoma is most likely in the scenario of over-drainage resulting in forward rotation of the ciliary body. It is one of the most complex and difficult of all the glaucomas to treat and it can progress to permanent blindness without prompt intervention. Eyes in which administration of atropine has been unsuccessful require urgent surgical intervention, which is beyond the scope of this article.

Wipe out

The phenomenon of severe visual loss after surgery, with no obvious cause, is known as 'wipe-out' or 'snuff syndrome'. Wipe-out may affect patients who have very severe glaucomatous damage and overall is a very uncommon complication but remains an important concern among glaucoma surgeons. Although, wipe-out was probably more common after full thickness filtration surgery, it is not known whether patients who had been diagnosed with wipe out had other undetected causes such as macular oedema, hypo-tonous maculopathy or inflammation. With modern surgical techniques this entity is becoming increasingly rare. Precautions such as changes in anesthesia techniques, careful monitoring of perioperative IOP spikes and careful attention to avoid both high and low postoperative pressures will help make this entity a thing of the past.

Conclusion

In conclusion, every surgeon, even with the most meticulous attention to correct surgical approach and technique, will at some stage encounter most of the complications mentioned here. Active interventions to avoid complications are therefore common, occurring in about half of post-operative patients at some stage. I hope the above is of some help in management strategies to improve our surgical results from this fundamental operation to prevent blindness from glaucoma. There are many other potential compli-cations of trabeculectomy during the first three months post-operatively that space does not permit me to explore.

Please remember: prevention is better than cure and "first do no harm"!

Ocular haemangioblastoma and renal cell carcinoma in von Hippel-Lindau disease – a case study

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The Journal of the College of Ophthalmologists of Sri Lanka 2016; 22: 81-86

Introduction

Von Hippel-Lindau syndrome (VHL) is a familial cancer predisposition syndrome that is characterized by visceral cysts and benign tumours in multiple organ systems that have subsequent potential for malignant change. This may include many different types of tumours such as haemangioblastoma, renal cell carcinoma (RCC), renal cysts and phaeochromocytoma (6).

The name originates from Eugene von Hippel and Arvid Lindau, who described retinal and other tumors in 1904 and 1926 respectively (7).

It is a rare disorder that affects approximately 1 in 36,000 live births (7).

VHL is caused by highly penetrant mutations in the VHL gene (3p25.3), a classic tumour suppressor (6).

This is an autosomal dominant disorder requiring only a single mutation to confer susceptibility. 20% of patients with this disorder have no family history and it is assumed that they acquired a new mutation (7).

There is over 90% penetrance by the age of 65. Age at diagnosis varies from infancy to age 60-70 years, with an average patient age at clinical diagnosis of 26 years (8).

Case report

A 23 years old patient presented with the complaint of pain and photophobia with gradual onset proptosis of his right eye with visual acuity of light perception. Examination showed a vascular mass lesion over superotemporal aspect of right globe with non-axial proptosis. Anterior segment showed evidence of band keratopathy, clotted hyphaema with high intraocular pressure. The lesion progressed very rapidly with increased vascularity within few weeks as shown in Figure 1 and 2.

He gave a significant past ocular history. He presented to the National Eye Hospital 10 years back, with redness of right eye following trauma with a tennis ball. Right eye visual acuity was 1/60 with relative apparent pupillary defect. Fundal examination showed superotemporal reddish mass lesion, surrounded exudates with detached retina (Figure 3). With the aid of fundal fluorescin angiogram (Figure 4), a retinal haemangioma was suspected and retinal cryotherapy was given. Thereafter the patient defaulted follow up due to lack of family support.

Figure 1.

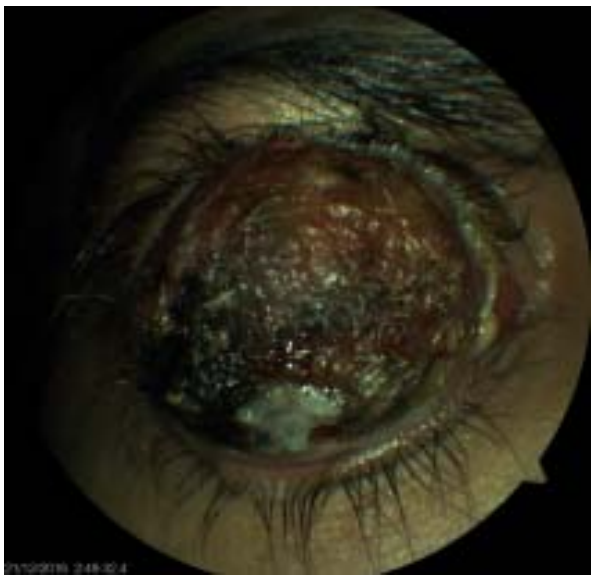

Figure 2.

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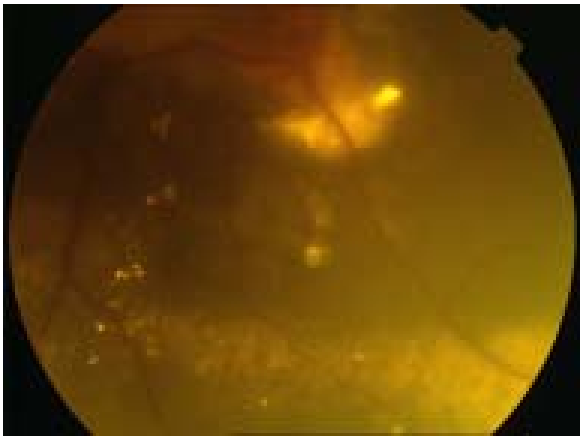

Figure 3.

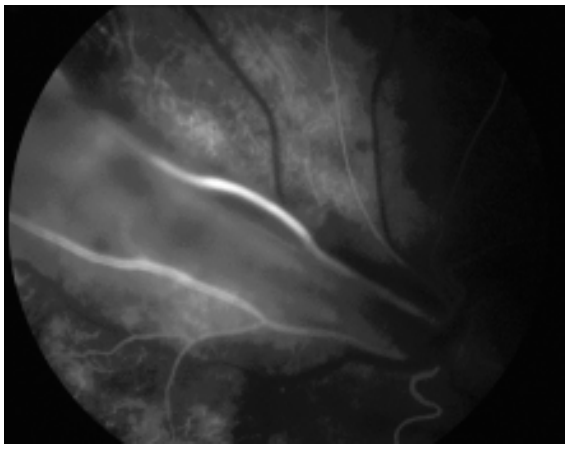

Figure 4.

Investigations

MRI of brain with MR angiography (Figure 5 and 6) showed enlarged right globe with nonhomogeneous signal and vascularity suspecting a vascular tumour or Haemangioma. Intraconal compartment appeared normal with prominant vascularity in the region. No intracranial extention noted.

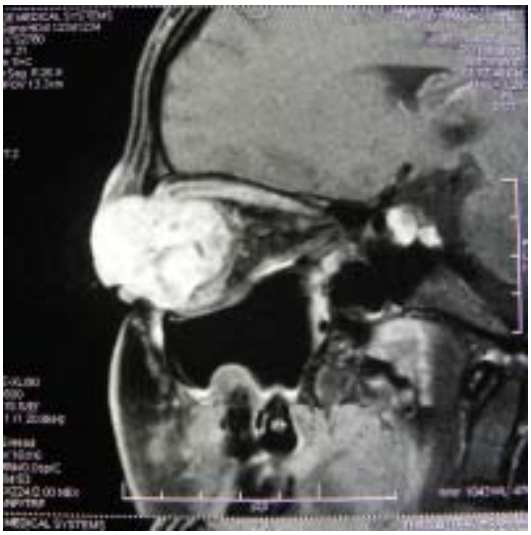

Figure 5.

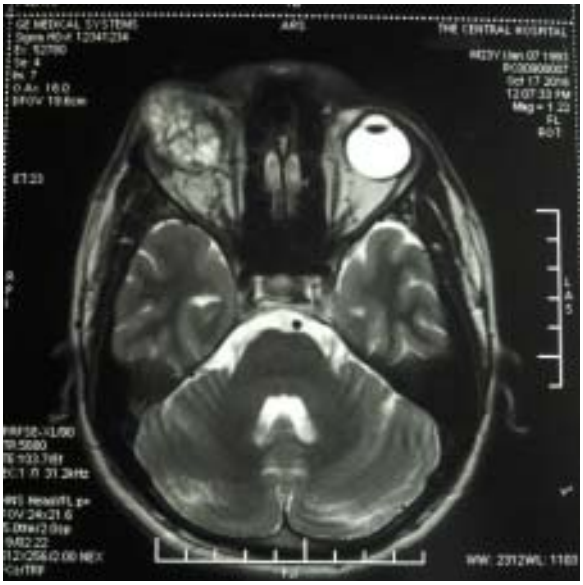

Figure 6.

Digital subtraction angiography (Figure 7) showed abnormal collection of vessels in right globe suggestive of vascular malformation.

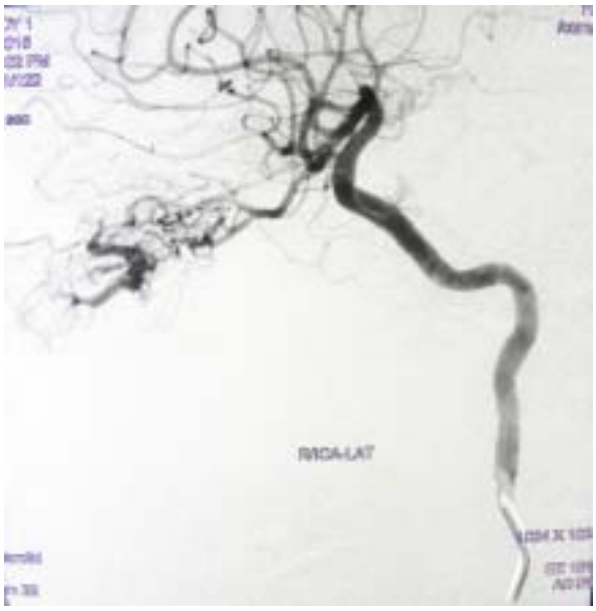

Figure 7.

The patient was referred to occuloplastic surgeon who advocated embolization to reduce the vascularity of proptosed eye. Following referral as shown in Figure 8, embolization of right ophthalmic artery was done by interventional radiologist at National Hospital of Sri Lanka.

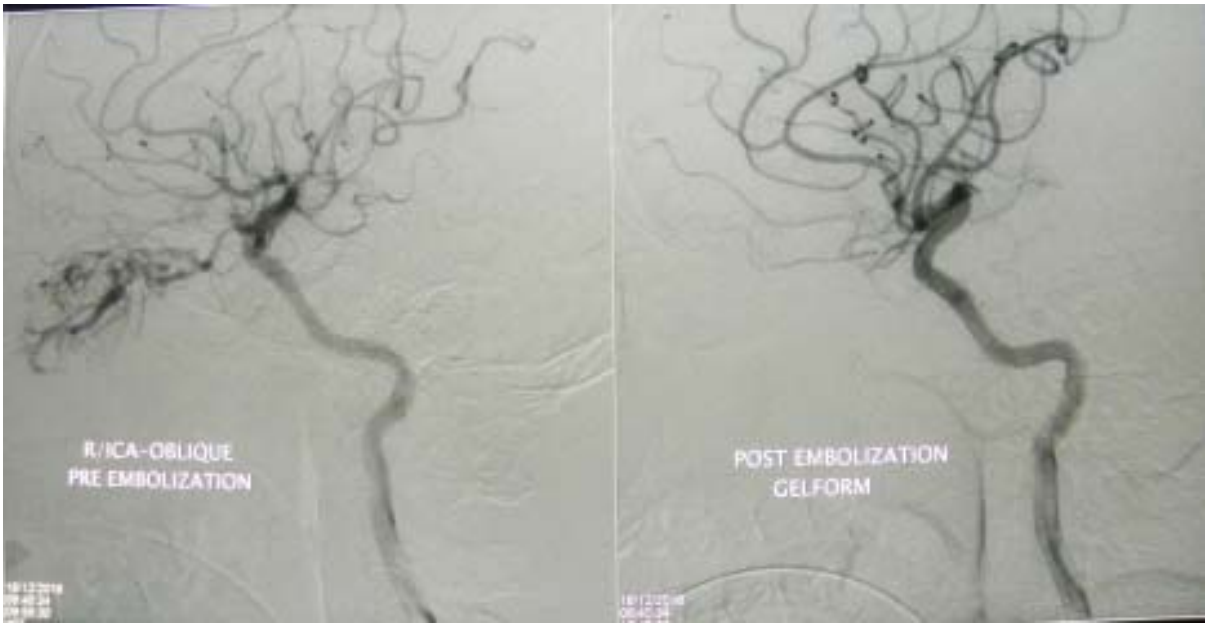

Figure 8.

Due to the possible anastomotic feeding vessels within orbit and the difficult in accessing the orbital apex due to the size of tumour, right sided craniotomy and enucleation was done by combined effort of consultant neurosurgeon and oculoplastic surgeon with minimum postoperative complications.

Histopathology revealed (Figure 9 and 10) abundant thin wall small vascular channels with numerous stromal cells consist of vaculated abundant clear cytoplasm and hyperchromatic nuclei suggestive of a WHO grade 1 haemangioblastoma of right eye.

The patient developed fever and abdominal pain 3 months after the enucleation and CT urogram was done at Colombo South Teaching Hospital.

The CT urogram (Figure 11 and 12) was reported as multiple bilateral renal cell carcinoma, all of them of $T_1N_0M_0$ grade and hence von Hippel-Lindau disease was suspected as the possible diagnosis.

Due to multiple tumours and the possible risk of haemangioblastoma of central nervous system, the contrast enhanced CT of brain was also done and no evidence of intracranial pathology. Left partial nephrectomy had been performed at Colombo South Teaching Hospital.

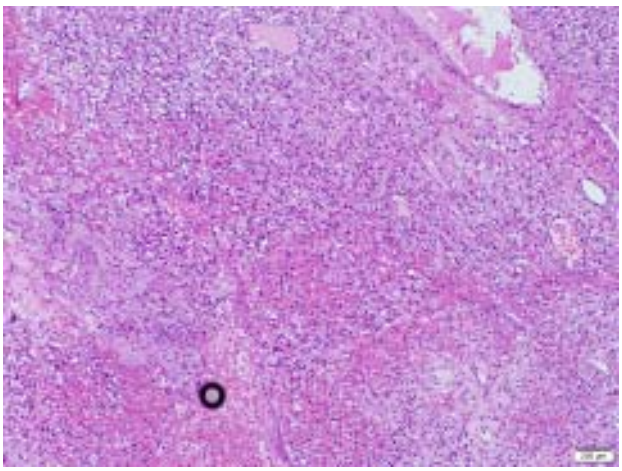

Figure 9.

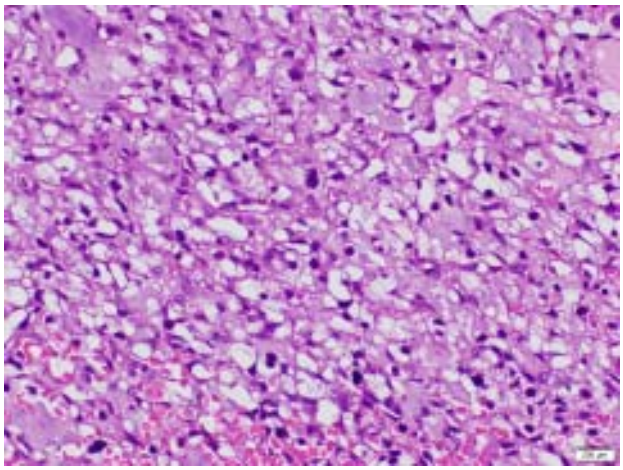

Figure 10.

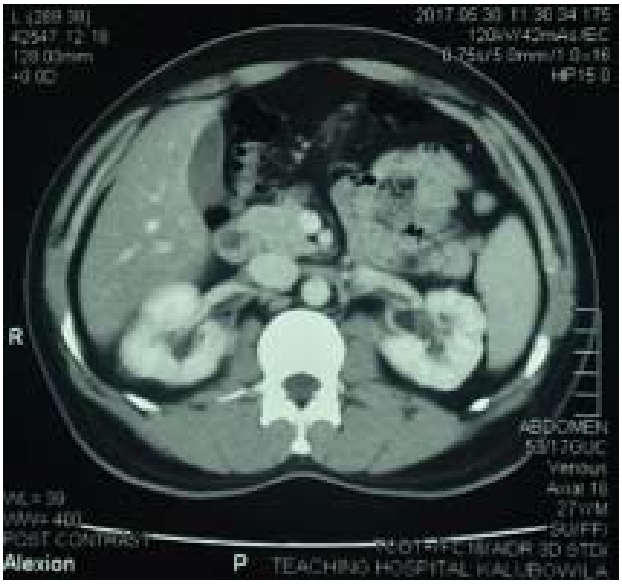

Figure 11.

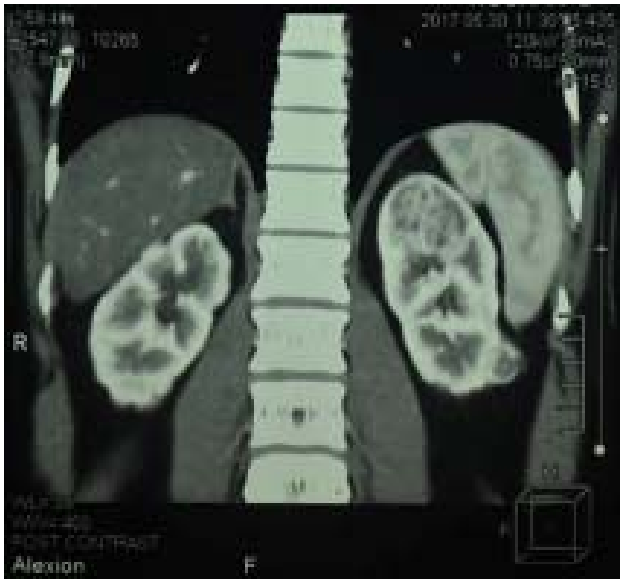

Figure 12.

Discussion

Because VHL can cause malignant tumours, it is considered one of a group of familial cancer risk diseases, which are transmitted genetically.

Features can be remembered by the mnemonic HIPPEL (2):

Haemangioblastoma

Increased risk of RCC

Phaeochromocytoma

Pancreatic lesions (cysts, **C**ystadenomas, cystadenocarcinomas)

Eye dysfunction (retinal haemangioblastomas)

Liver, renal and pancreatic cysts

Disease features

CNS haemangioblastomas affect approximately two-thirds of patients with the disorder, making them the most common type of tumour. Although benign, they can cause significant morbidity depending on their location in the CNS (7).

Retinal haemangiomas diagnosed in 50% of patients with VHL disease, these haemangiomas are composed of endothelial cells and pericytes. These are usually supplied by large dilated feeder vessels, may occur in any part of the retina. Serum leakage from these vessels and haemangiomas leads to retinal exudates. Organized fibroglial bands with traction retinal detachment and vitreous haemorrhage may occur, along with potential complications such as glaucoma or permanent vision loss (5).

Phaeochromocytomas are usually benign but may cause symptoms such as headaches, palpitations and episodes of shock-like signs (7).

RCC and cysts affect up to 60% of patients and are usually asymptomatic (7).

Pancreatic tumours are usually asymptomatic (7).

Differential diagnoses

Multiple endocrine neoplasia, neurofibromatosis, polycystic kidney disease, tuberous sclerosis, Birt-Hogg-Dube syndrome, and hereditary pheochromocytoma-paraganglioma syndromes associated with succinate dehydrogenase subunit mutations (SDHB, SDHC and SDHD) (6).

Diagnosis

Because VHL disease is a multiple-organ disease that widely varies in clinical presentation, various manifestations may lead to diagnosis. Criteria are the following (8):

- More than one haemangioblastoma in the CNS (brain, spinal cord) or eye.
- A single haemangioblastoma in the CNS or retina, plus a visceral manifestation (multiple renal, pancreatic, or hepatic cysts; pheochromocytoma; renal cancer).
- Positive family history plus any one of the above manifestations.
- Elucidation of a deleterious mutation in VHL gene.

This patient having a haemangioblastoma of right eye with multiple renal cysts, he fulfilled the second criteria to diagnose as VHL disease. A detailed family history revealed his mother has passed away when he was 3 years and other facts that are suspicious of a positive family history. We are planning to do genetic studies to elucidate the mutation of VHL gene at the genetic laboratory of University of Colombo with help of Prof.Vajira Dissanayake.

Techniques such as southern blotting and gene sequencing can be used to analyse DNA and identify mutations. de novo cases that produce genetic mosaicism are more difficult to detect because mutations are not found in the white blood cells that are used for genetic analysis (1,3).

Antenatal diagnosis is possible through molecular analysis of amniocytes or chorionic villus cells if a disease-causing mutation has been identified in an affected family member (6).

Treatment

Early diagnosis of VHL syndrome is preferable to allow timely management of the disorder and reduce the impact on the life expectancy. Previously, life expectancy was approximately 50 years, but this has improved significantly with the introduction of screening and early management (7).

For this reason, individuals with VHL disease are usually screened routinely for retinal angiomas, CNS haemangioblastomas, clear-cell renal carcinomas and pheochromocytomas (4).

We have done CT brain to exclude CNS haemangioblastomas in our patient and his CT abdomen didn't show any features of pheochromocytomas as well.

Standard therapies

Retinal haemangioblastomas

Small peripheral lesions can be successfully treated with little to no loss of vision using laser. Larger lesions often require cryotherapy as for our patient in his first presentation at the age of 13. If the lesion left untreated, as in our patient who defaulted his treatment, the lesion spreads slowly within years and may require enucleation with careful haemostasis. If the haemangioblastoma is on the optic disc, there are few treatment options that will successfully preserve vision. The optimal treatment would be a drug, but to date none has proven successful (10).

Renal cell carcinoma

A strategy for ensuring that an individual will have a sufficiently functioning kidney throughout his or her lifetime begins with careful monitoring and choosing to operate only when tumour size or rapid growth rate suggest the tumour may gain metastatic potential (at approximately 3 cm).

This patient had bilateral involvement of his kidneys at the time of diagnosis, mainly of left side. So the technique of kidney-sparing surgery such as partial nephrectomy of left side was done.

Radio frequency ablation (RFA) or cryosurgery (cryotherapy) may be considered as to the right kidney. especially for smaller tumours at earlier stages. Care must be taken not to injure adjacent structures and to limit scarring which may complicate subsequent surgeries (10).

Brain and spinal haemangioblastomas

Symptoms related to haemangioblastomas in the brain and spinal cord depend on tumour location, size, and the presence of associated swelling or cysts. Symptomatic lesions grow more rapidly than asymptomatic lesions. Cysts often cause more symptoms than the tumour itself. Once the lesion has been removed, the cyst will collapse. If any portion of the tumour is left in place, the cyst will re-fill. Small haemangioblastomas which are not symptomatic and are not associated with a cyst have sometimes been treated with stereotactic radiosurgery, but this is more a preventive than a treatment, and long-term results seem to show only marginal benefit (10).

Pancreatic neuroendocrine tumours

Careful analysis is required to differentiate between serious cystadenomas and pancreatic neuroendocrine tumours (pancreatic NETs). Cysts and cystadenomas generally do not require treatment. Pancreatic NETs should be rated on size, behaviour, and DNA type (10).

Pheochromocytomas

Surgical removal is performed after adequate blocking with medication, and laparoscopic partial adrenalectomy is preferred. Vital signs are carefully monitored for at least a week following surgery while the body readjusts to its "new normal". Special caution is warranted during surgical procedures of any type and during pregnancy and delivery. Even pheochromocytomas that do not appear to be active or causing symptoms should be removed, ideally prior to pregnancy or non-emergency surgery (10).

Prognosis

Prognosis depends on the occurrence of multiple tumours. RCC is the main cause of death, followed by CNS haemangioblastomas. Average life expectancy was

previously estimated to be 50 years; however, regular surveillance and early detection and management of tumours have now reduced the morbidity and mortality (6).

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