"THE OCCURRENCE OF KERATOCONUS(KC) IN DOWN SYNDROME(DS) "

Dr Anitha Venugopal

Professor

Cornea and Refractive services

Aravind Eye Care System
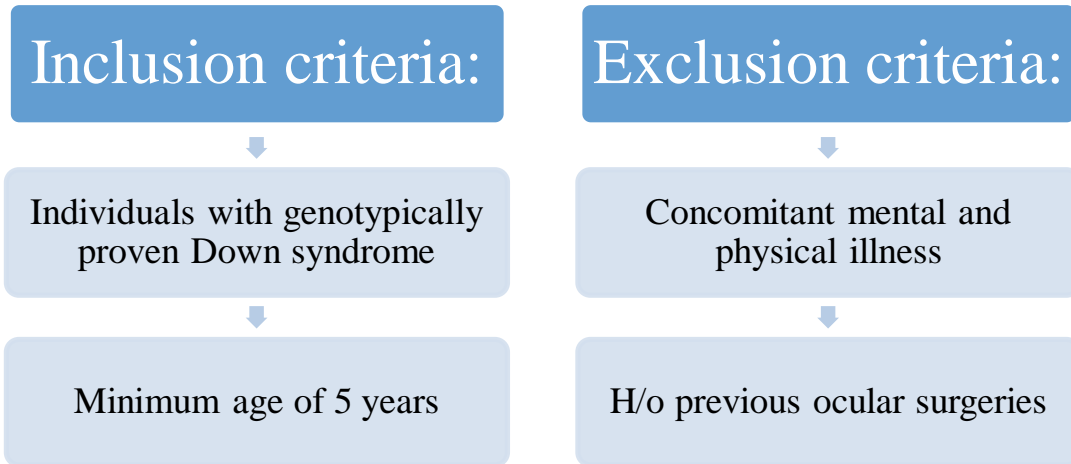
Tirunelveli, Tamil Nadu, India

Introduction

➢ Keratoconus (KC) is a progressive corneal thinning and ectasia leading to visual impairment.

➢ The prevalence of KC in general population is 1 in 2300 per one lakh in central India (0.003% -2.3%).

➢ The prevalence in DS ranges from 0% to 71%, which is about 30% more than in general population.

➢ DS individuals present late with severe visual impairment due to advanced stage of the disease and visual loss due to inability in communicating their symptoms with care givers.

Purpose

➢ To assess the occurrence of Keratoconus using diagnostic tomographic parameters in Down syndrome individuals

Materials & Methods

✓ A Cross- sectional observational study

✓ The Down syndrome individuals were enrolled through screening at tertiary eye care center, Tamil Nadu, India from special needs schools, and relevant non-profit organizations.

✓ Total 65out of 69 individuals with Down syndrome were included for the study

Inclusion criteria:	Exclusion criteria:
Individuals with genotypically proven Down syndrome	Concomitant mental and physical illness
Minimum age of 5 years	H/o previous ocular surgeries

Methodology

- ✓ Slit lamp examination
- ✓ Visual acuity testing using Snellen acuity chart
- ✓ Pentacam imaging done by a skilled optometrist
- ✓ Imaging repeated until acceptable quality acquired
- ✓ If > 3 attempts needed, another appointment scheduled

Tomographic parameters:
Z Kmax ART max
I-S value
BAD-D value
Mean CT
ARC, PRC PPI
AE/PE
Anterior COMA/ Posterior COMA HOA

Definitions

➤Diagnosis of KC was made independently by two experienced cornea specialists

Clinical criteria

Munson's sign
Vogt's striae
Fleischer's ring

Para clinical criteria

Kmax >48.0D
ART max<339um
I-S value>1.4D
BAD-D >1.6

Mean CT<490
ARC<7.05mm
PRC<5.70mm
PPI>0.13
AE/PE>2um/>5um
Ant/Post coma 0.05$\pm$0.17
HOA >0.34$\pm$0.08

Results

- 20.8% diagnosed as KC

- KC manifests more in females 92.3%

- Mean age of KC and KC suspects was 13.46 and 12.52 years.

Table1: Demographic characters of the downs syndrome patients based on diagnosis					
	Advanced KC	KC	KCS	FFKC	No KC
Number of eyes, n	8(6.1)	27(20.8)	61(46.9)	5(3.8)	29(22.3)
Age, years					
Mean (SD)	21.00(8.89)	13.46(7.50)	12.52(7.57)	20.75(8.06)	16.33(6.43)
Min - Max	11 to 28	3 to 31	4 to 40	13 to 32	8 to 33
Gender, n (%)					
Male	2(66.7)	1(7.7)	20(66.7)	2(50.0)	9(60.0)
Female	1(33.3)	12(92.3)	10(33.3)	2(50.0)	6(40.0)
Eye, n (%)					
Right eye	3(37.5)	13(48.1)	30(49.2)	4(80.0)	15(51.7)
Left eye	5(62.5)	14(51.9)	31(50.8)	1(20.0)	14(48.3)
Scissor reflex, n (%)					
Yes	2(25.0)	5(20.0)	1(1.7)	0	0
No	6(75.0)	20(80.0)	58(98.3)	5(100.0)	29(100.0)
Corneal scar, n (%)					
Yes	3(37.5)	0	0	0	0
No	5(62.5)	17(100.0)	48(96.0)	5(100.0)	24(100.0)
Sub epithelial haze	0	0	2(4.0)	0	0

Table2: Corneal topography details of the patients by diagnosis

	Diagnosis, median(interquartile range)					P-value
	Advanced KC	KC	KCS	FFKC	No KC	
ZK max	93.4(60.1, 102.0)	52.2(51.3, 54.2)	49.10(47.7, 50.1)	47.4(46.9, 47.9)	46.7(46.1, 47.58)	<0.001
ART max	0.5(0.0, 2.0)	138.0(69.0, 247.0)	279.0(223.0, 346.0)	303.0(266.0, 440.0)	361.0(273.0, 409.0)	<0.001
I-S value	2.1(-1.1, 5.5)	1.5(0.1, 3.1)	0.7(-0.2, 1.8)	-0.1(-0.3, 1.0)	0.4(-0.1, 1.0)	0.184
BADD	37.5(9.2, 73.0)	4.6(2.7, 7.7)	2.4(1.6, 3.4)	1.5(1.1, 1.6)	1.6(1.3, 2.6)	<0.001
Mean CT	230.5(159.5, 345.0)	446.0(415.0, 479.0)	480.0(471.0, 504.0)	504.0(487.0, 504.0)	496.0(479.0, 523.0)	<0.001
ARC	6.8(5.8, 7.6)	6.8(6.7, 7.3)	7.2(7.1, 7.6)	7.3(7.3, 7.4)	7.5(7.3, 7.6)	<0.001
PRC	4.8(3.6, 6.1)	5.4(5.3, 5.8)	5.9(5.7, 6.2)	6.1(6.1, 6.2)	6.2(6.0, 6.3)	<0.001
PPI	1.3(0.0, 59.5)	1.7(1.1, 3.4)	1.2(1.0, 1.7)	1.1(1.1, 1.2)	1.0(0.8, 1.2)	0.003
AE	-39.0(-298.0, 20.0)	1.0(-4.0, 9.0)	3.0(-1.0, 6.0)	2.0(-2.0, 3.0)	4.0(0.0, 5.0)	0.561
PE	81.0(14.0, 112.0)	16.0(7.0, 45.0)	7.0(3.5, 12.0)	3.0(0.0, 4.0)	7.0(2.0, 11.0)	0.001
ANT coma	-0.4(-1.6, 1.1)	-0.4(-1.0, 0.1)	-0.1(-0.5, 0.1)	-0.2(-0.2, -0.0)	-0.1(-0.3, -0.0)	0.496
Post coma	0.3(-0.1, 1.8)	0.1(-0.1, 0.1)	0.04(-0.04, 0.1)	-0.04(-0.1, -0.03)	-0.04(0.0, 0.1)	0.372
HOA	3.3(1.3, 8.2)	1.3(0.8, 2.0)	1.0(0.6, 1.5)	0.5(0.3, 0.6)	0.6(0.5, 0.7)	<0.001

Mean values of tomographic indices such as Z Kmax, ART max, BAD-D, Mean CT, ARC, PRC and HOA are noted to be statistically significant between various diagnosis of KC.

Table3: Tabulation of DS patient's visual acuity

VA classification	Diagnosis, n (%)					KC group VA
	Advanced KC	KC	KCS	FFKC	No KC	
6/6 to 6/12	3(37.5)	4(14.8)	18(29.5)	0	17(58.6)	25(24.8)
6/18 to 6/36	1(12.5)	6(22.2)	15(24.6)	1(20.0)	1(3.4)	23(22.8)
<6/36	2(25.0)	1(3.7)	4(6.6)	0	0	7(6.9)
Not co-operative	2(25.0)	16(59.3)	24(39.3)	4(80.0)	11(37.9)	46(45.5)
Total	8	27	61	5	29	101
Mean (SD)	0.63 (0.37)	0.63 (0.67)	0.44 (0.35)	0.48	0.13 (0.20)	0.49 (0.43)
Median	0.54	0.48	0.48	0.48	0.00	0.48
IQR	0.30 to 1.00	0.30 to 0.60	0.18 to 0.60	-	0.00 to 0.18	0.18 to 0.60

➢ Z K max and HOA are observed to be the best discriminants to predict KC from Non KC groups in DS population.

➢ There was a significant difference in the average visual acuity between the diagnoses

Table4: ROC results between DS-KC and No KC

Index	AUROC	Cutoff	Sensitivity (%)	Specificity (%)
ZKmax	0.83	48.80	69.7	96.6
ART max	0.51	513.00	1.0	100.0
I-S value	0.63	1.05	46.0	79.0
BAD-D	0.69	1.62	83.0	55.0
Mean CT	0.53	538.00	6.0	100.0
ARC	0.57	7.80	13.1	100.0
PRC	0.54	6.63	8.8	100.0
PPI	0.69	1.22	50.0	88.9
AE	0.57	8.00	18.0	97.0
PE	0.61	21.00	22.0	100.0
Anterior vertical coma	0.61	0.06	28.0	93.0
Posterior vertical coma	0.60	0.11	27.0	93.0
HOA	0.74	0.87	59.0	90.0

Discussion

✓ Variable rates of prevalence and Incidence of KC in DS reported in literature

✓ 2 reasons: a) Cornea structure is different in DS individuals

✓ b) Lack of standard criteria for KC diagnosis in DS

Fair discriminants: K max & Corneal thickness
Better Diagnostic indices: IVA, I-S value, IHD, ISV
Highest diagnostic indices: Anterior HOA, Total HOA, AVC, PVC

➢ Thinner, steeper corneae, thinner crystalline lenses, reduced accommodative function, and variations in tear film composition.

➢ An increased prevalence of high astigmatism, keratoconus, and cataract.

➢ These variations in the optical and refractive components of the DS eye may influence optical quality and integrity and help to explain the poor visual performance found in this unique group.

Mapping the corneal thickness and volume in patients with Down syndrome: a comparative population-based study

Hassan Hashemi [1], Ali Makateb [2], Shiva Mehravaran [3], Akbar Fotouhi [4], Fereshteh Shariati [1], Soheila Asgari [1]

Best KC discrimators: HOA and Coma

For KCS: Minimum CT, Corneal volume and BAD-D

Third order Vertical Coma

⬇

Fair diagnostic index for KCS

⬇

Excellent for KC

Eye and Vision

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Tomography-based definition of keratoconus for Down syndrome patients

Soheila Asgari[1], Shiva Mehravaran[2], Mohammadreza Aghamirsalim[3] and Hassan Hashemi[1*]

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Total coma measured by OPD scan	Fair discriminant for FFKC	Excellent for diagnosis of KC

✓ Our study also in agreement with the previous studies: HOA and Z Kmax were considered best predictive indices for KC in DS

Limitations

- ✓ Small Sample size

- ✓ Pilot study- follow-up and evaluation not done

- ✓ Sub group analysis not done

- ✓ Comparison not done with normal subjects with and without KC

Conclusion

- ✓ The occurrence of KC in our study participants was 20.8%, quite high

- ✓ All DS individuals need to be subjected to tomographic screening for early detection of KC

- ✓ Recommended to promptly manage them with CXL to stop the progression of the disease

- ✓ To improve the visual quality of life