



**Moorfields**  
Eye Hospital Dubai

A BRANCH OF MOORFIELDS LONDON

## Irfan Khan

**DRCOphth, MRCOphth, FRCOphth, PGME, CCST UK**  
**Consultant Paediatric Ophthalmologist**



a place where I can be a child

**SickKids®**



Edge Hill University



**M R E H 2 0 0**

MANCHESTER ROYAL EYE HOSPITAL 1814-2014  
TWO HUNDRED YEARS OF EXCELLENCE



**Moorfields**  
Eye Hospital Dubai

A BRANCH OF MOORFIELDS LONDON

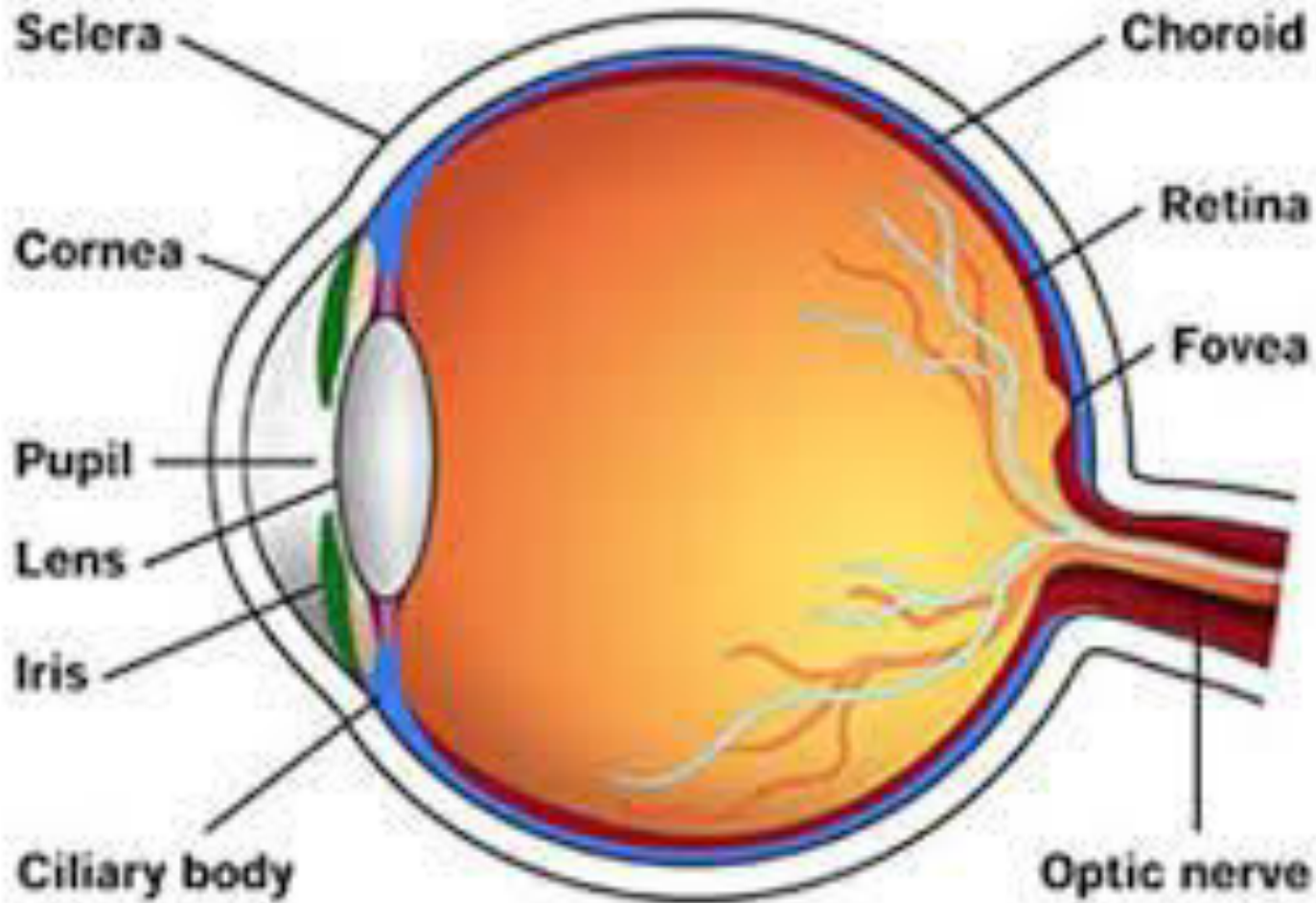
# **Diagnosis made or missed in a blink of a eye ! Ophthalmic Clinical Scenarios**

**Kids Heart Pediatric Conference**

**Al Ain**

**March 2018**

[irfan.khan@moorfields.ae](mailto:irfan.khan@moorfields.ae)





**LOGARITHMIC VISUAL ACUITY CHART "HOTV"**

FOR TESTING AT 400M (16 INCHES)  
SIZE INCREMENTS - 0.1 LOG UNITS

LETTER SIZE	ORIGINAL		SMELLEN DISTANCE EQUIVALENT METER FOOT	DIOPTERS OF ADD FOR 1 M
6.0 M	.060	H O V T O	6/100 20/400	20 D
5.3 M	.063	O V H V T	6/90 20/320	18 D
4.8 M	.068	T H V O H	6/75 20/250	13 D
4.0 M	0.70	H O T V O	6/60 20/200	10 D
3.2 M	.083	V T H O V	6/48 20/160	8 D
2.5 M	.085	T O V H O	6/36 20/125	6 D
2.0 M	.090	V H O T V	6/30 20/100	3 D
1.6 M	.095	O T H V O	6/24 20/80	4 D
1.25 M	.098	H O V T V	6/18 20/60	3 D
1.0 M	.099	T H O H	6/15 20/50	2.5 D
80 M	0.80		6/12 20/40	
63 M	0.63		6/9.0 20/30	
50 M	0.50		6/7.5 20/25	
40 M	0.40		6/6 20/20	
30 M	0.30		6/4.5 20/15	
25 M	0.25		6/3.6 20/12.5	
20 M	0.20		6/3 20/10	

Precision Vision®  
300 First Street - La Salle, IL 61830 - U.S.A. - Phone 815 226-3000 - FAX 815 224-2224  
email: precisionvision@precisionvision.com • www.precisionvision.com

CAT. NO. 2004



# Fixing and Following Light



Moorfields  
Eye Hospital Dubai  
A BRANCH OF MOORFIELDS LONDON



07/05/2018

irfan.khan@moorfields.ae

# Object to Occlusion of Eye



Moorfields  
Eye Hospital Dubai  
A BRANCH OF MOORFIELDS LONDON



# Direct Ophthalmoscope



Moorfields  
Eye Hospital Dubai  
A BRANCH OF MOORFIELDS LONDON





# Red Reflex



Moorfields  
Eye Hospital Dubai  
A BRANCH OF MOORFIELDS LONDON



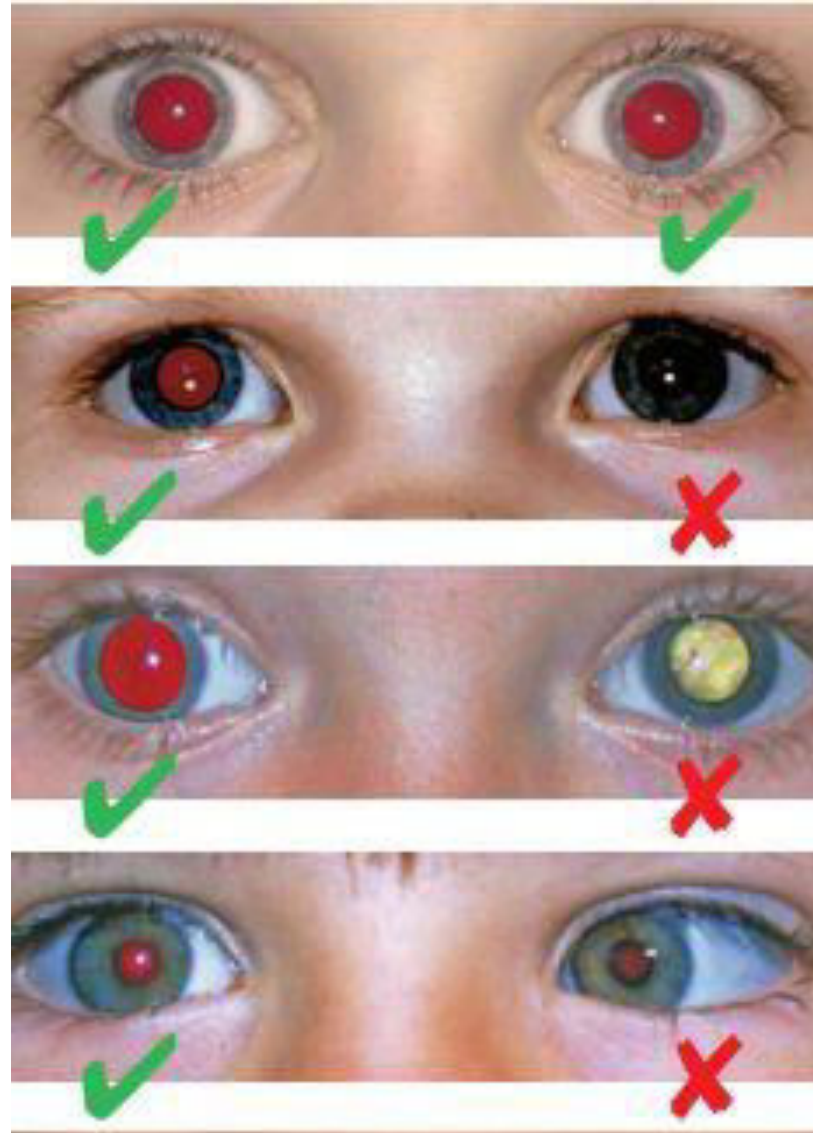
07/05/2018

irfan.khan@moorfields.ae

# Red Reflex



Moorfields  
Eye Hospital Dubai  
A BRANCH OF MOORFIELDS LONDON



07/05/2018

irfan.khan@moorfields.ae



Baby MP

East African family

Gestation age 27/40

Birth weight 800 gms

Screening for Retinopathy of Prematurity



- Fetal bradycardia
- Absent end diastolic flow
- Oligohydramnios
- Emergency LSCS for severe IUGR
- Cardiac massage & Adrenaline
- Intubation & ventilation
- Surfactant
- Extubated day 4
- Antibiotics for sepsis
- FFP clotting derangements

# Ophthalmic examination



Moorfields  
Eye Hospital Dubai  
A BRANCH OF MOORFIELDS LONDON



07/05/2018

irfan.khan@moorfields.ae







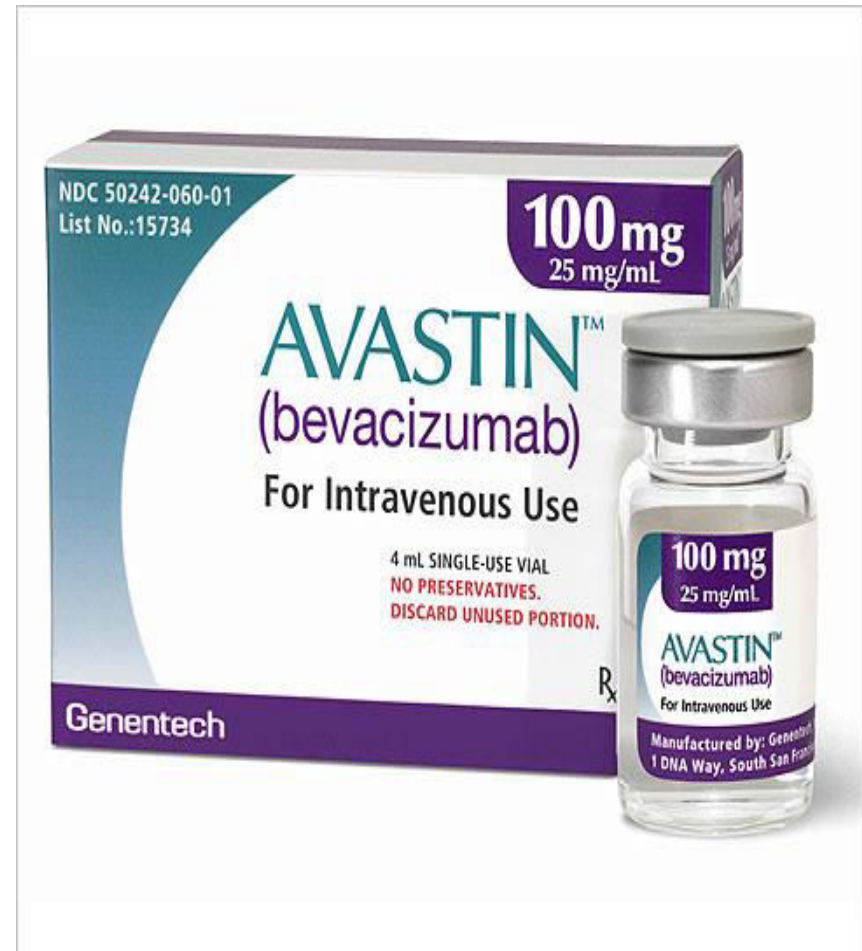
Laser Photocoagulation

or

Intra vitreal Anti Vegf



0.75mg of  
avastin pars plana  
Under anaesthesia

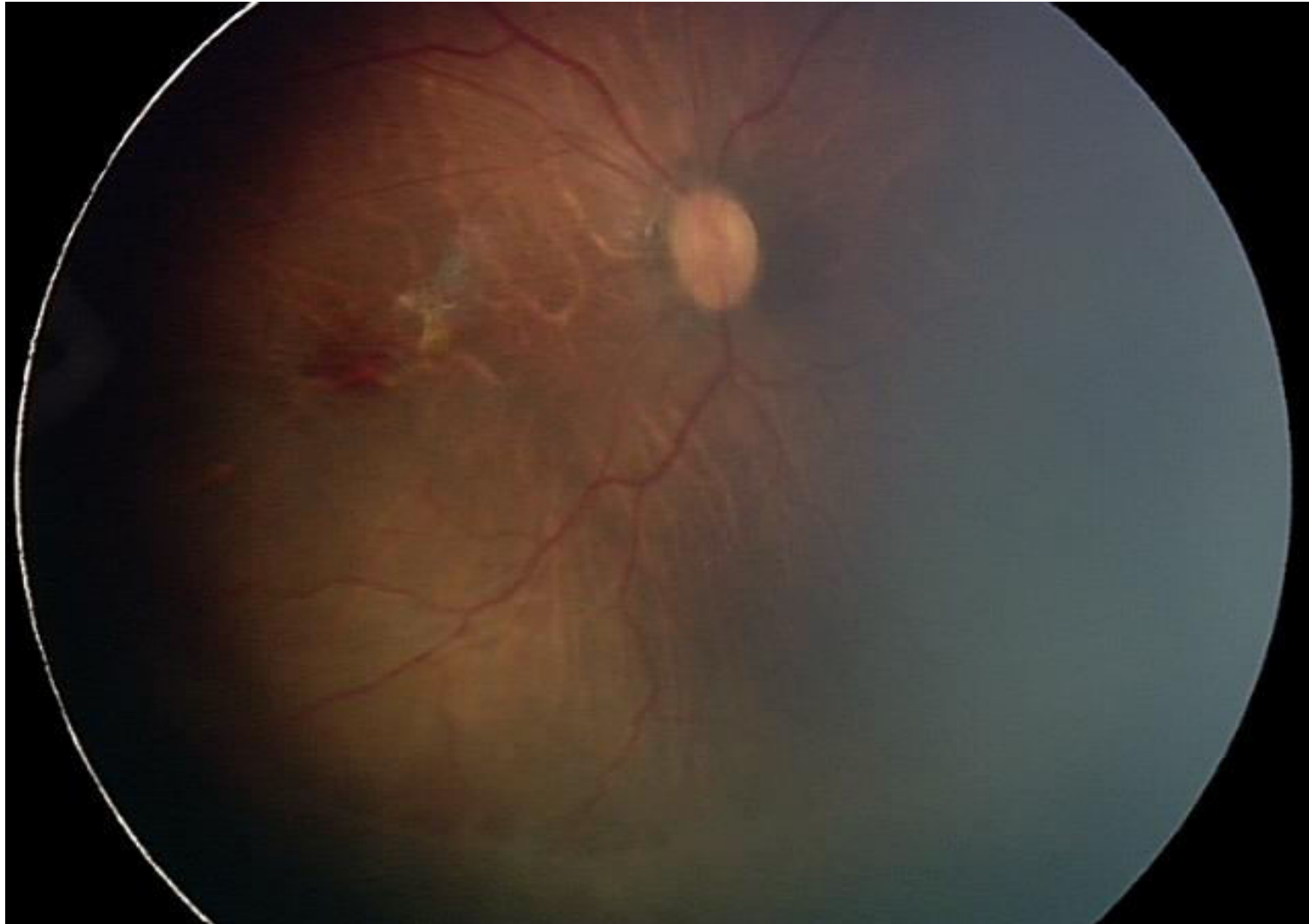




# 6 days Post Anti vegf

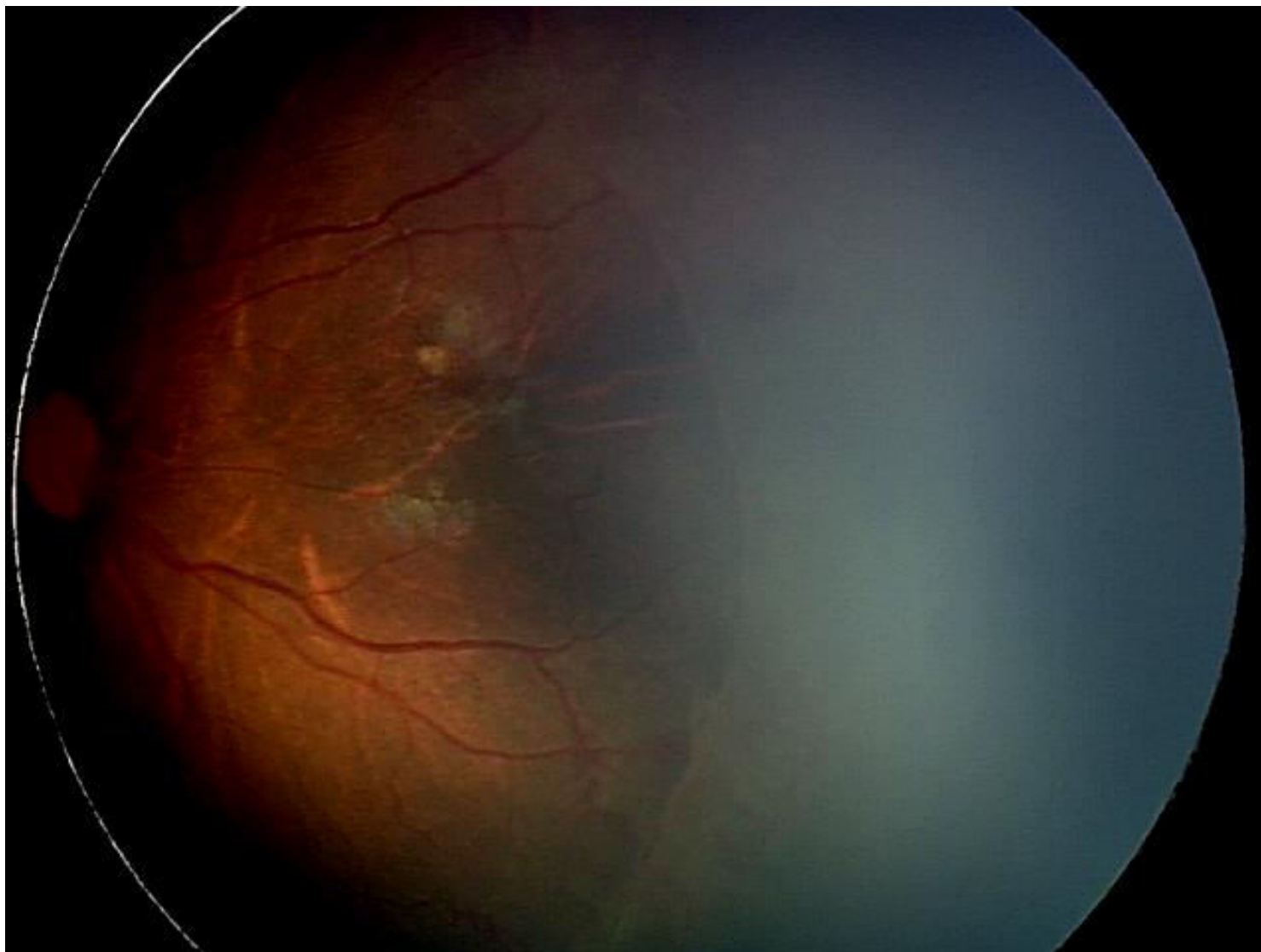


Moorfields  
Eye Hospital Dubai  
A BRANCH OF MOORFIELDS LONDON



07/05/2018

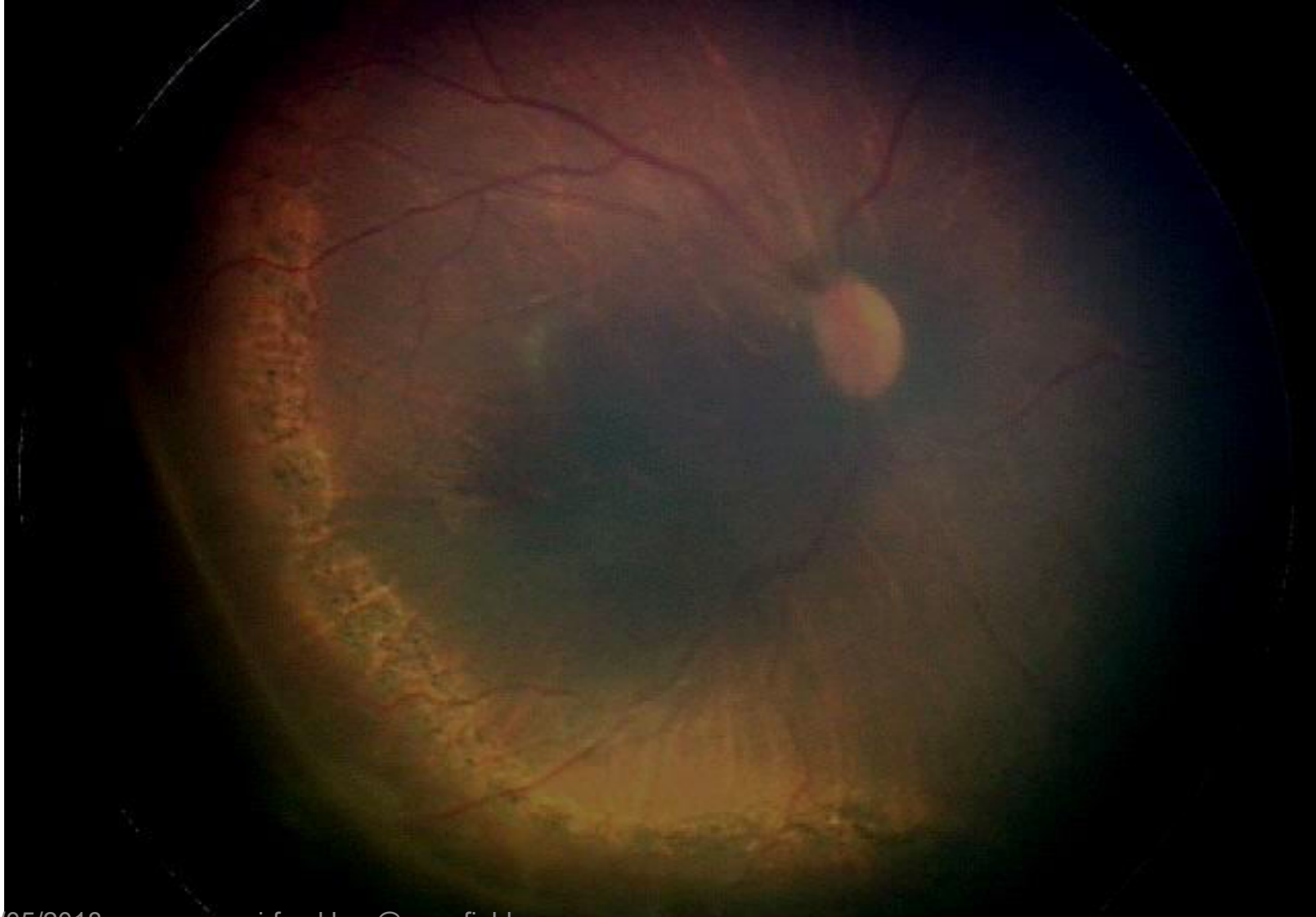
irfan.khan@moorfields.ae



# 6 days post Diode Laser



Moorfields  
Eye Hospital Dubai  
A BRANCH OF MOORFIELDS LONDON



07/05/2018

irfan.khan@moorfields.ae

# 2 weeks post Diode



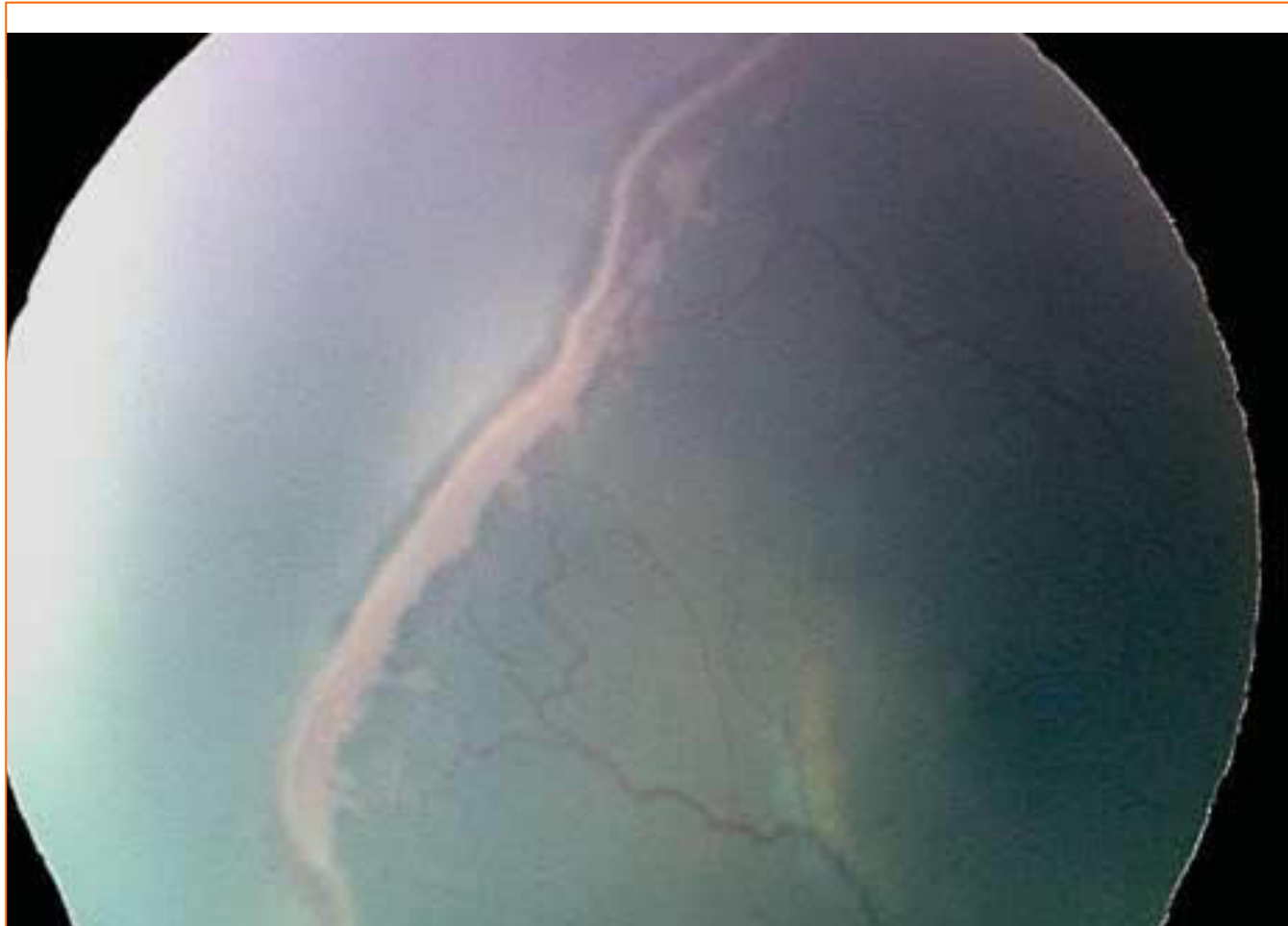
Moorfields  
Eye Hospital Dubai  
A BRANCH OF MOORFIELDS LONDON



07/05/2018

irfan.khan@moorfields.ae







# Case 2



Moorfields  
Eye Hospital Dubai  
A BRANCH OF MOORFIELDS LONDON



07/05/2018

irfan.khan@moorfields.ae

# Case 3- Hyperthyroidism



Moorfields  
Eye Hospital Dubai  
A BRANCH OF MOORFIELDS LONDON



6 year old double vision in extremes of gaze  
Autoimmune hepatitis and Hyperthyroidism  
Lid Retraction -Lid Lag- Restriction of ocular movements-  
enlargement of muscle sparing the TENDONS





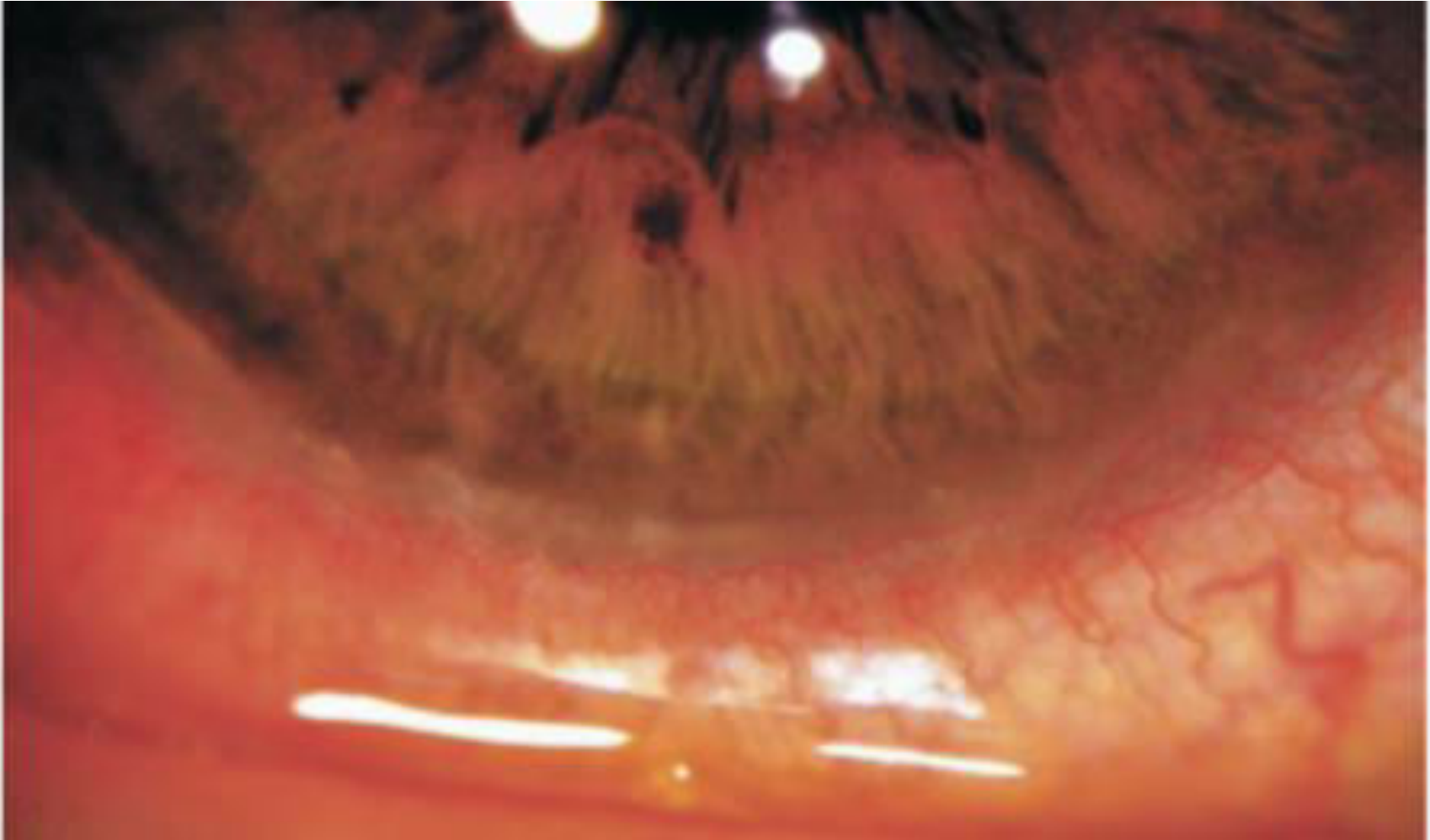
- 2.5% of Graves disease occur in children
- Second most common cause of proptosis in a series from Sickkids Toronto
- Well recognised in neonates of a hyperthyroid mother
- Rare before puberty
- Age of onset 12year older
- More common in girls 6:1
- Associations with diabetes and Downs Syndrome



# Case 3- Wegeners Granulomatosis



Moorfields  
Eye Hospital Dubai  
A BRANCH OF MOORFIELDS LONDON



07/05/2018

irfan.khan@moorfields.ae



- Necrotizing granulomatous vasculitis
- Airways and kidneys
- Under the age of 18
- Before Cyclophosphamide death in 2yrs
- Proptosis with bilateral pain
- ENT symptoms – nasal blockage, discharge, pain over sinuses, loss of hearing and tinitus

# Case 4



Moorfields  
Eye Hospital Dubai  
A BRANCH OF MOORFIELDS LONDON



07/05/2018

irfan.khan@moorfields.ae

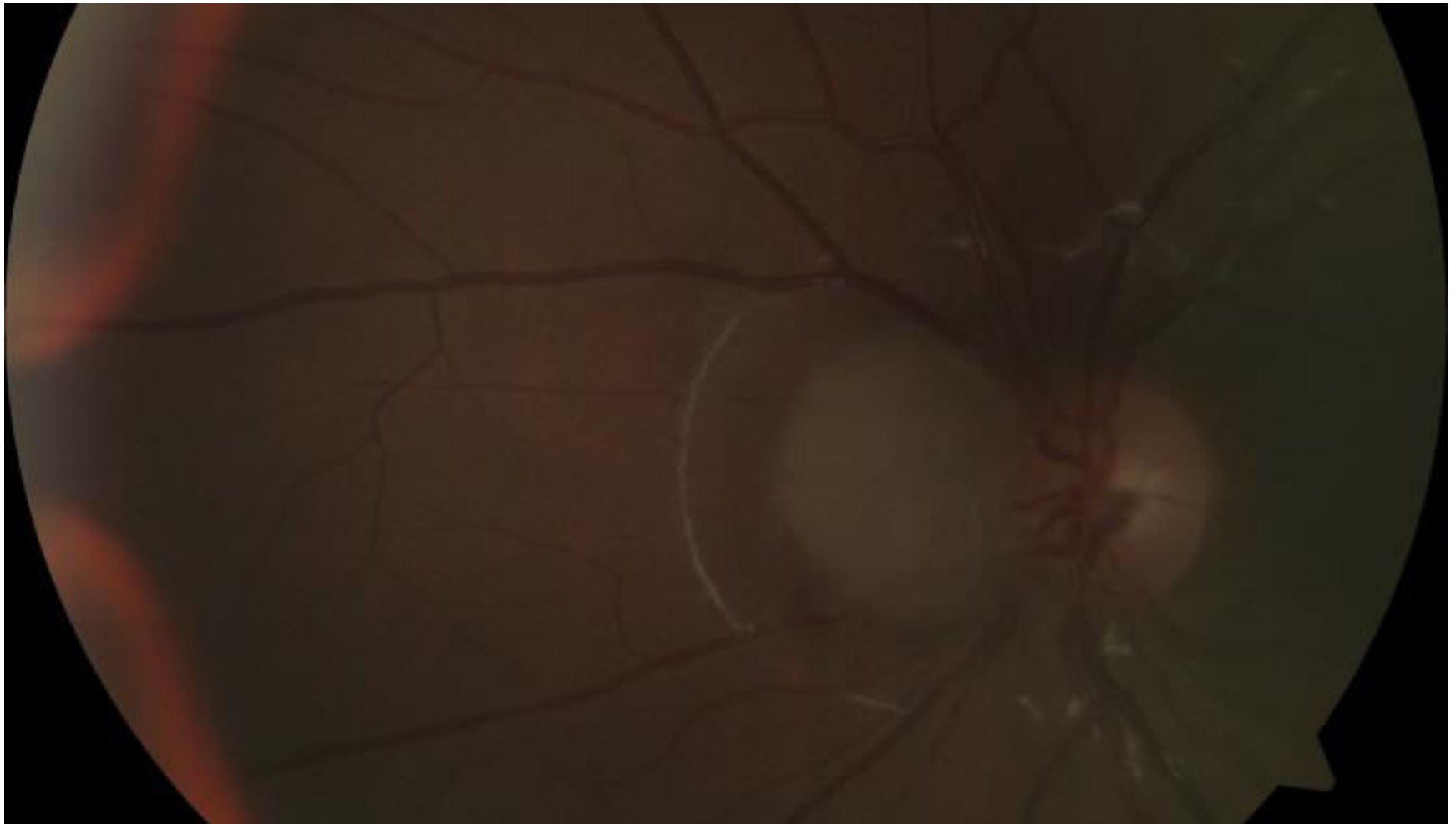


07/05/2018

irfan.khan@moorfields.ae



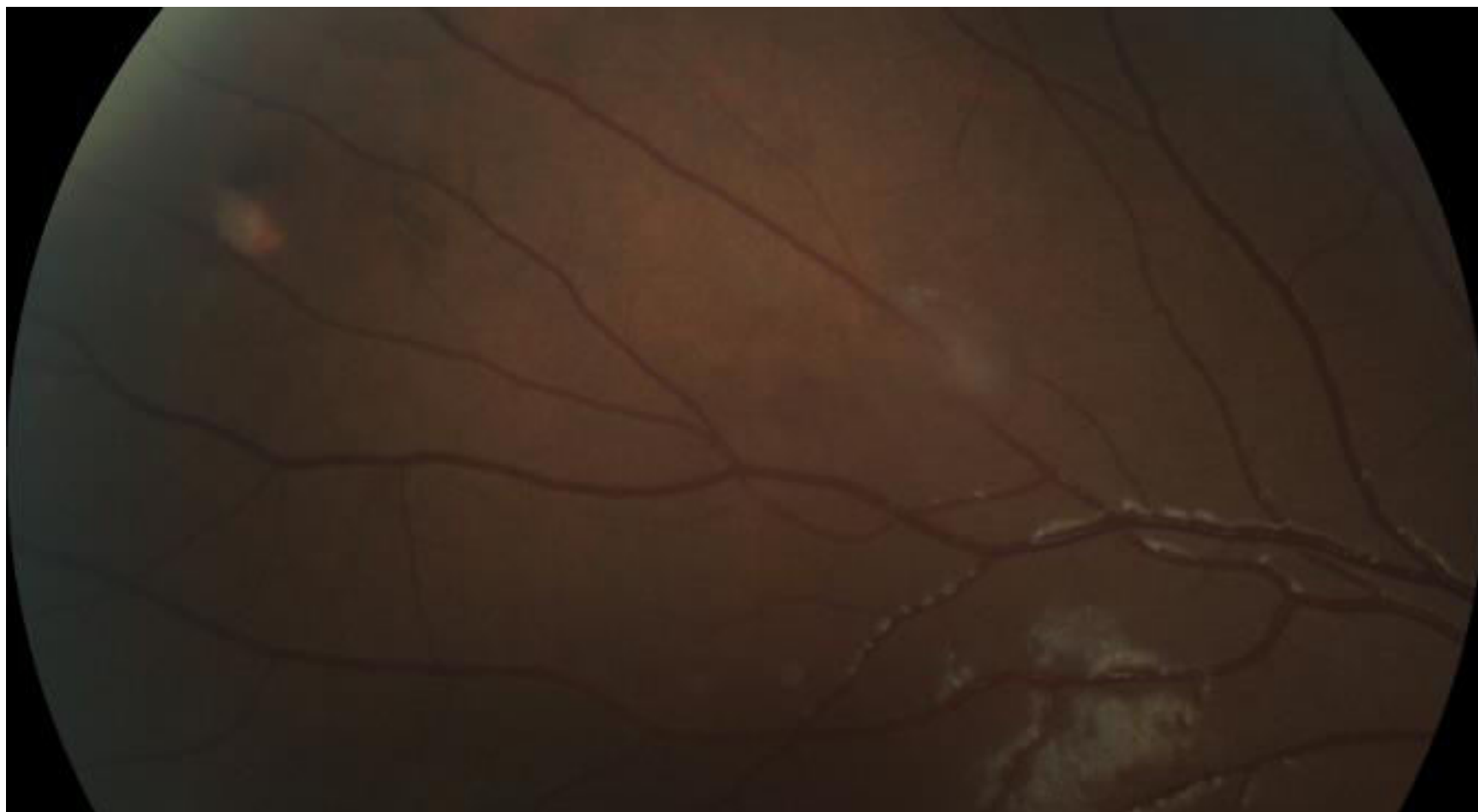


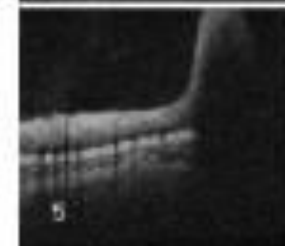
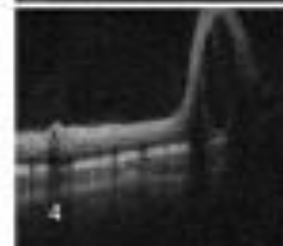
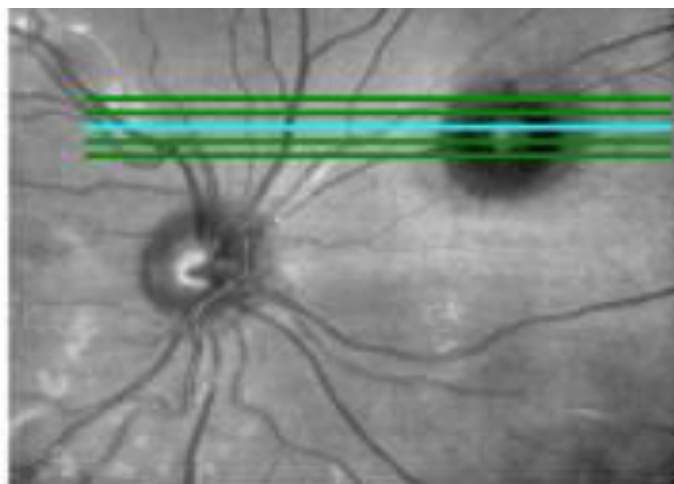


07/05/2018

irfan.khan@moorfields.ae









- Multisystem disorder
- Hamartomatous growth – Brain, Skin, Kidneys, eyes and heart
- Malignant tumors rare in kidney
- Presentations with epilepsy, developmental delay & learning difficulties

# CASE 5



Moorfields  
Eye Hospital Dubai  
A BRANCH OF MOORFIELDS LONDON

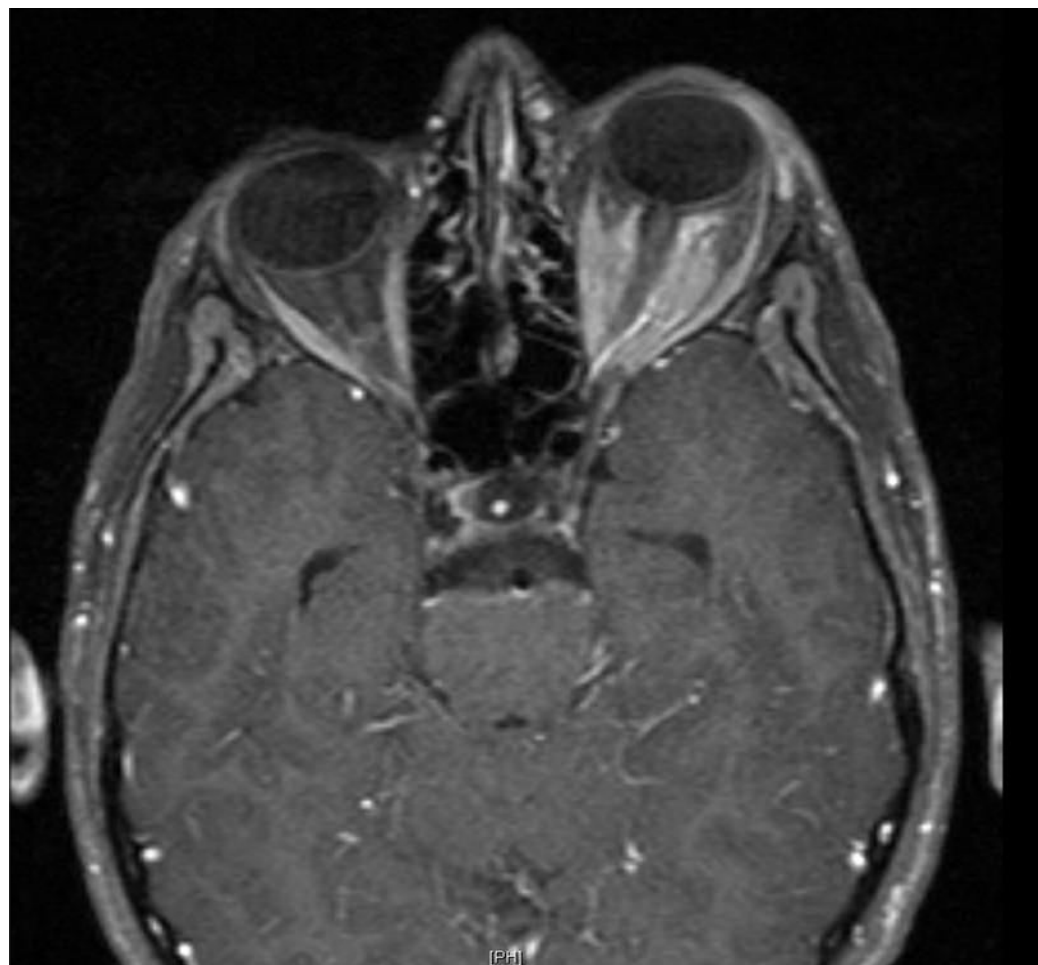


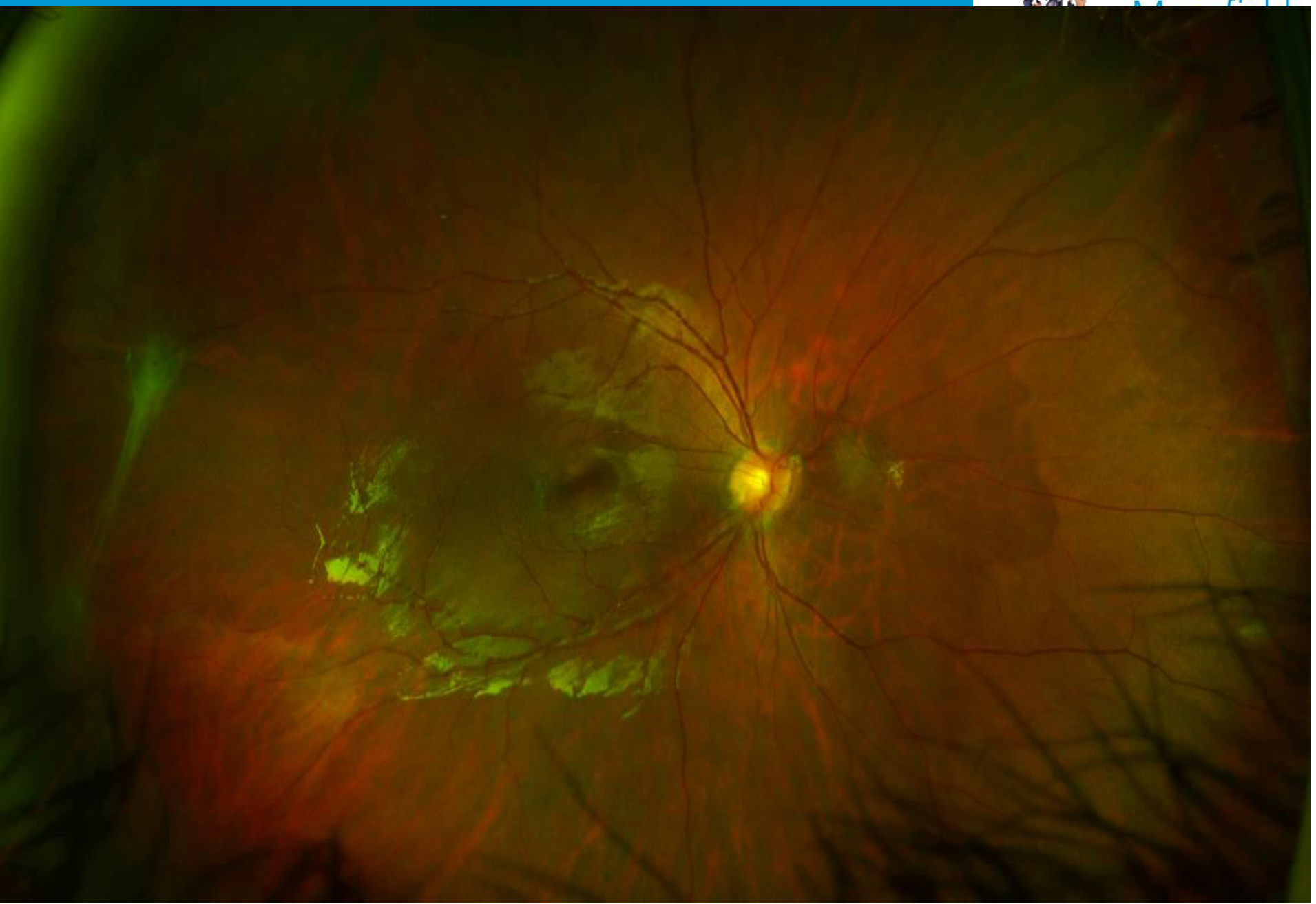






- 8 year old
- Referred for Proptosis for a year
- Vision 6/6
- Diplopia in left lateral gaze
- How to proceed





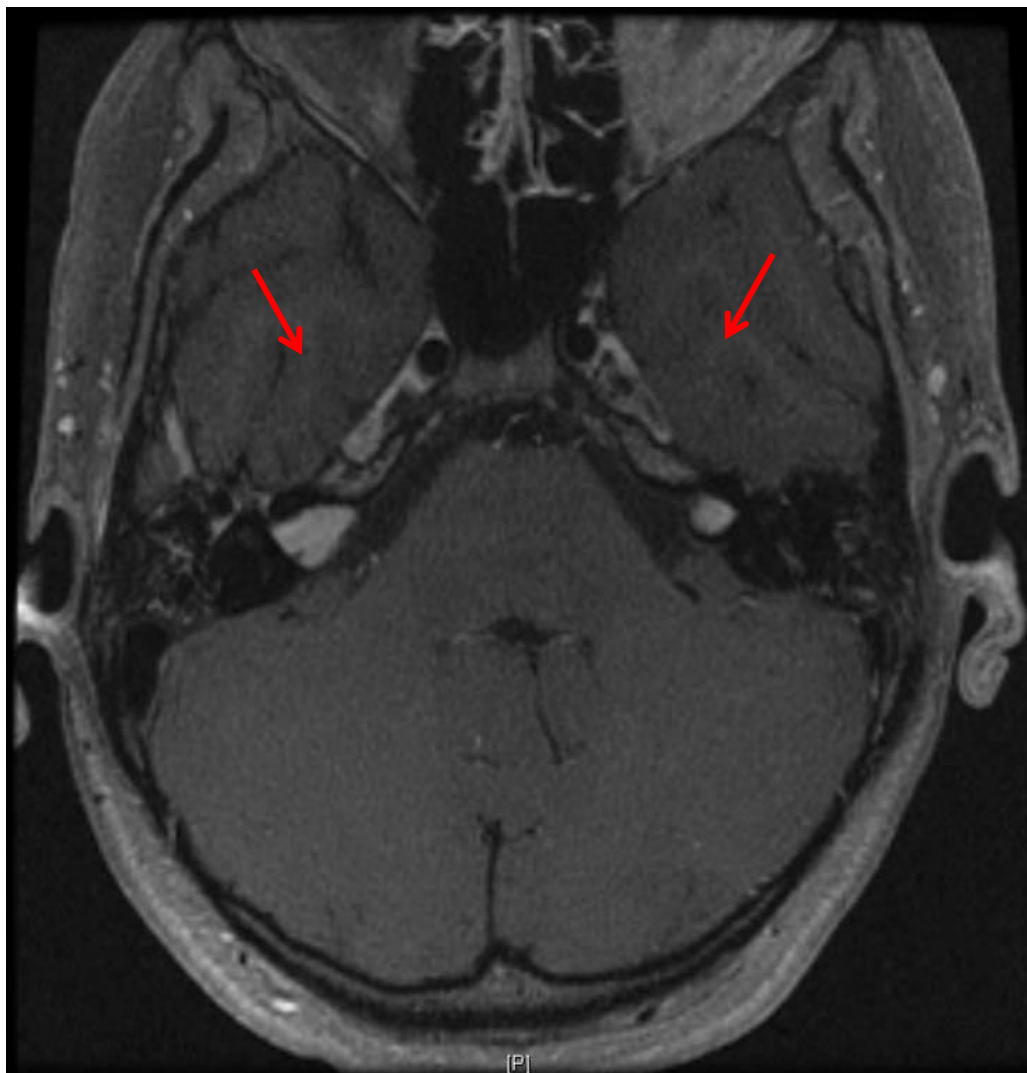




- Left orbital mass
- Left ERM
- Focal retinal lesions OU
- Asymptomatic



- Enlargement and enhancement of cisternal portions of trigeminal nerves bilaterally, extending into Meckel's cave (L>R)



- Enhancing masses in internal auditory canals (R>L) in keeping with small vestibular schwannomas



## Café-au-lait macules





# Left optic nerve sheath meningioma WITH

- 1) Bilateral trigeminal & vestibular schwannomas
- 2) Bilateral retinal astrocytic hamartomas
- 3) Multiple café-au-lait macules

Neurofibromatosis



Audiogram  
normal

Genetic testing for neurofibromatosis type 2

Screening MRI spine





- Left paraspinal mass T1-T6
- Abuts descending aorta, left CCA, left subclavian artery



- 8 year old
- Sudden onset decrease in vision
- VAR PL VAL 6/6
- Anterior segment unremarkable
- Few cells in the vitreous
- Uveitis work up
- History
- Cat Owner

# CASE 6

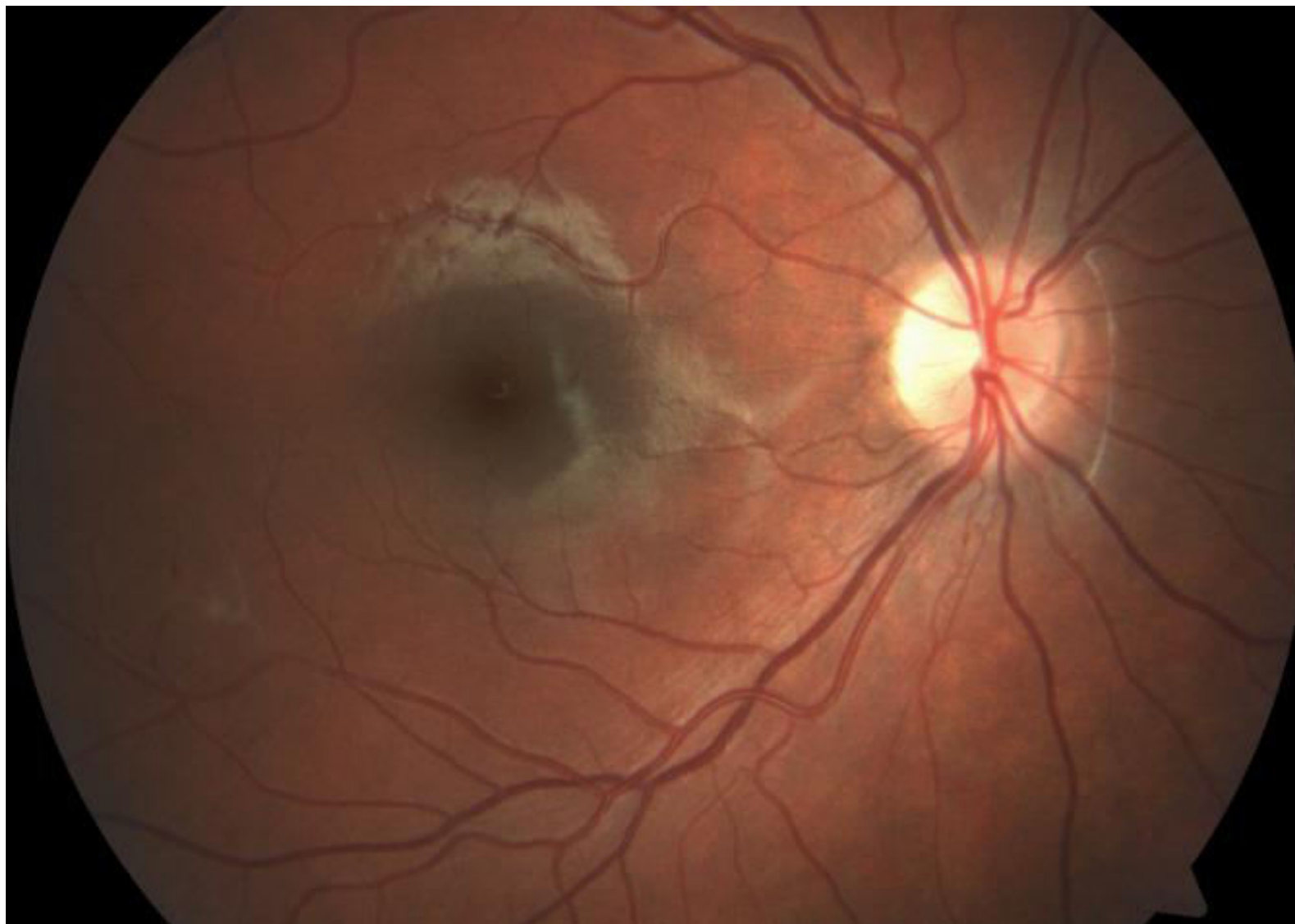


Moorfields  
Eye Hospital Dubai  
A BRANCH OF MOORFIELDS LONDON



07/05/2018

irfan.khan@moorfields.ae



# Bartonella- Cat Scratch Disease



Moorfields  
Eye Hospital Dubai  
A BRANCH OF MOORFIELDS LONDON



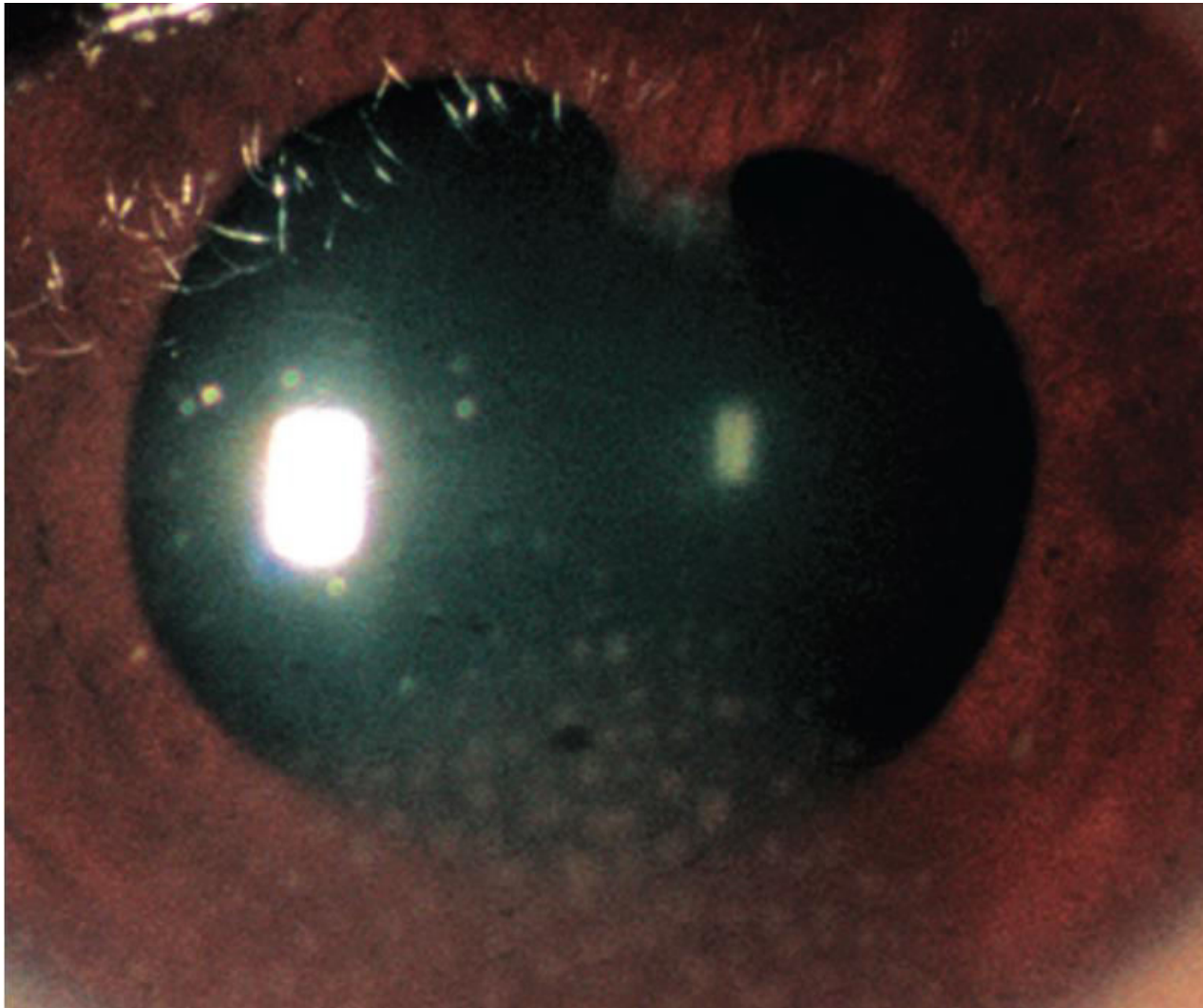
07/05/2018

irfan.khan@moorfields.ae

# Case 7



Moorfields  
Eye Hospital Dubai  
A BRANCH OF MOORFIELDS LONDON



07/05/2018

irfan.khan@moorfields.ae





# Juvenile Idiopathic Arthritis



Moorfields  
Eye Hospital Dubai  
A BRANCH OF MOORFIELDS LONDON



07/05/2018

irfan.khan@moorfields.ae



- Chronic joint inflammation starting before 16 yrs and lasting more than 6 weeks
- Incidence of JIA is 10/1000000
- Systemic onset JIA involves the eye in 1-2%
- Psoriasis is associated with ERA, IBD and Bechets.

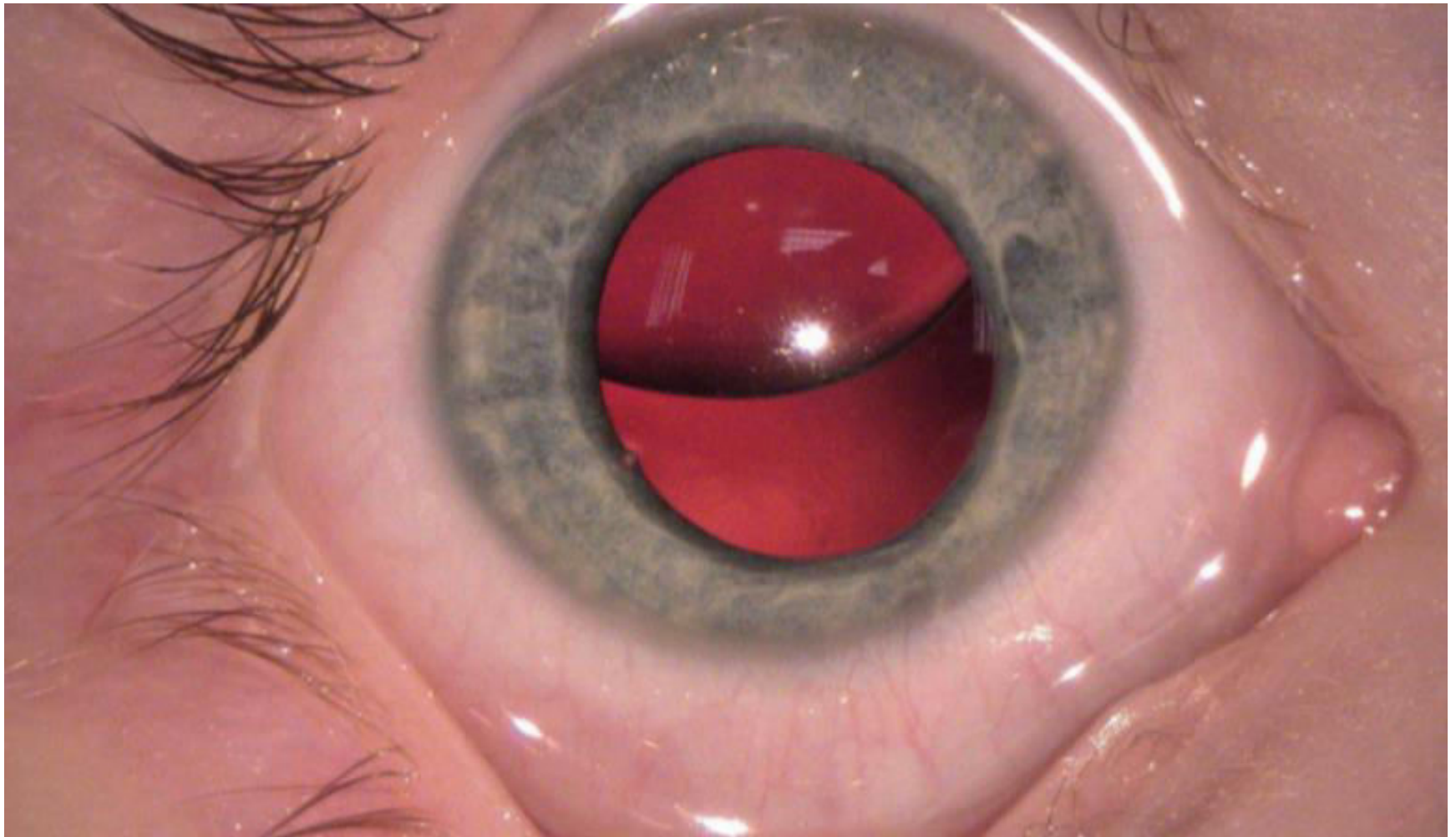


- Children developing arthritis below the age of 3yrs remain at risk for 7 years
- Those who develop arthritis after the age of 6 years are at risk for 3 years
- Development of uveitis after the age of 13yrs is rare
- Screening for uveitis - 3 months

# Case 8



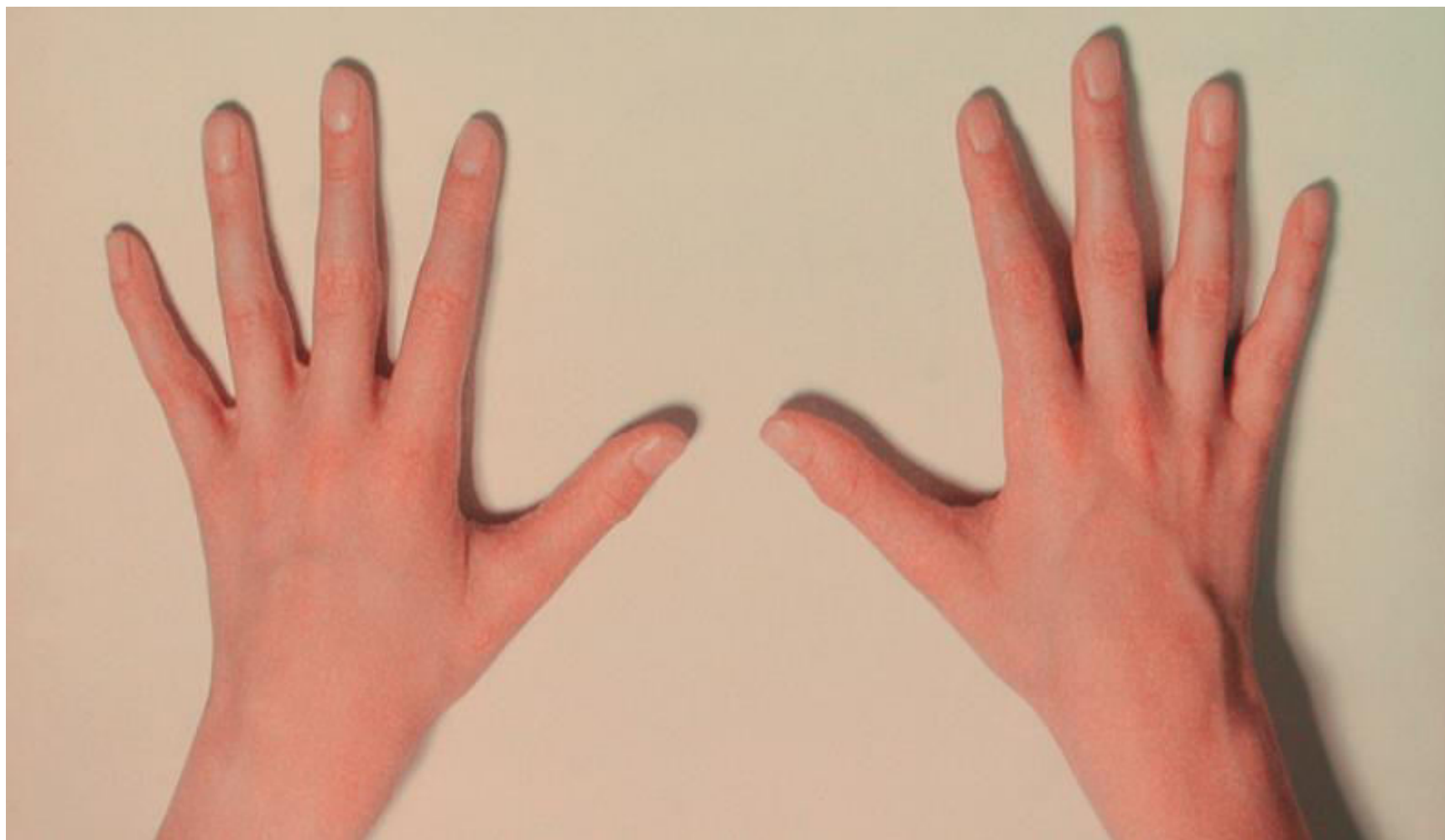
Moorfields  
Eye Hospital Dubai  
A BRANCH OF MOORFIELDS LONDON



07/05/2018

irfan.khan@moorfields.ae









- Mutation in FBN1
- Severe Neonatal MFS fatal by 2yrs
- Simple Ectopia lentis not associated with systemic diseases
- Chest wall deformities- dilatation of aortic root-mitral valve prolapse-aortic aneurysm
- Ectopia lentis 60%- Axial myopia-Corneal flattening-cataract-glaucoma and Strabismus
- Mostly displaced upwards

# Case 9

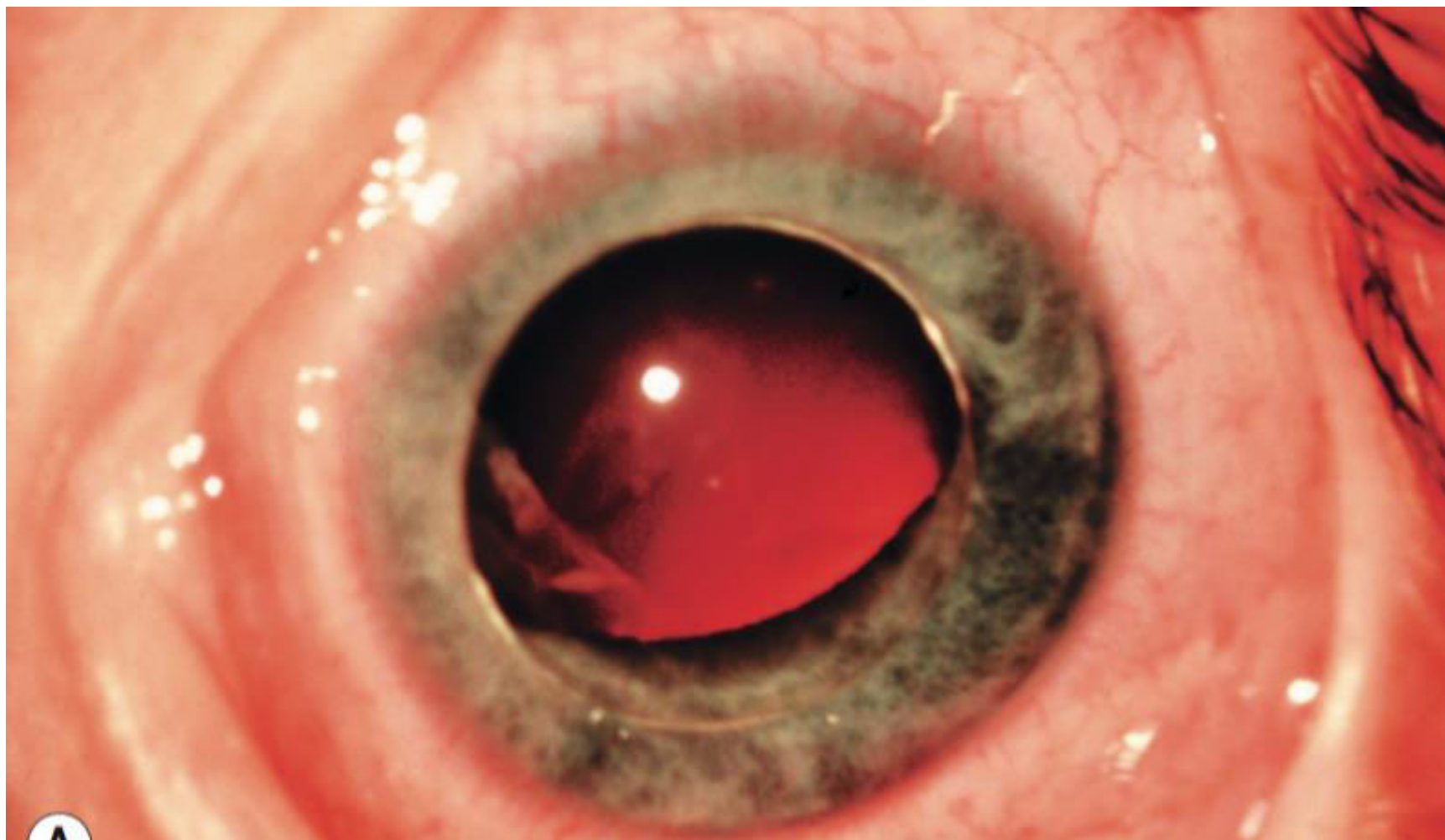


Moorfields  
Eye Hospital Dubai  
A BRANCH OF MOORFIELDS LONDON



07/05/2018

irfan.khan@moorfields.ae



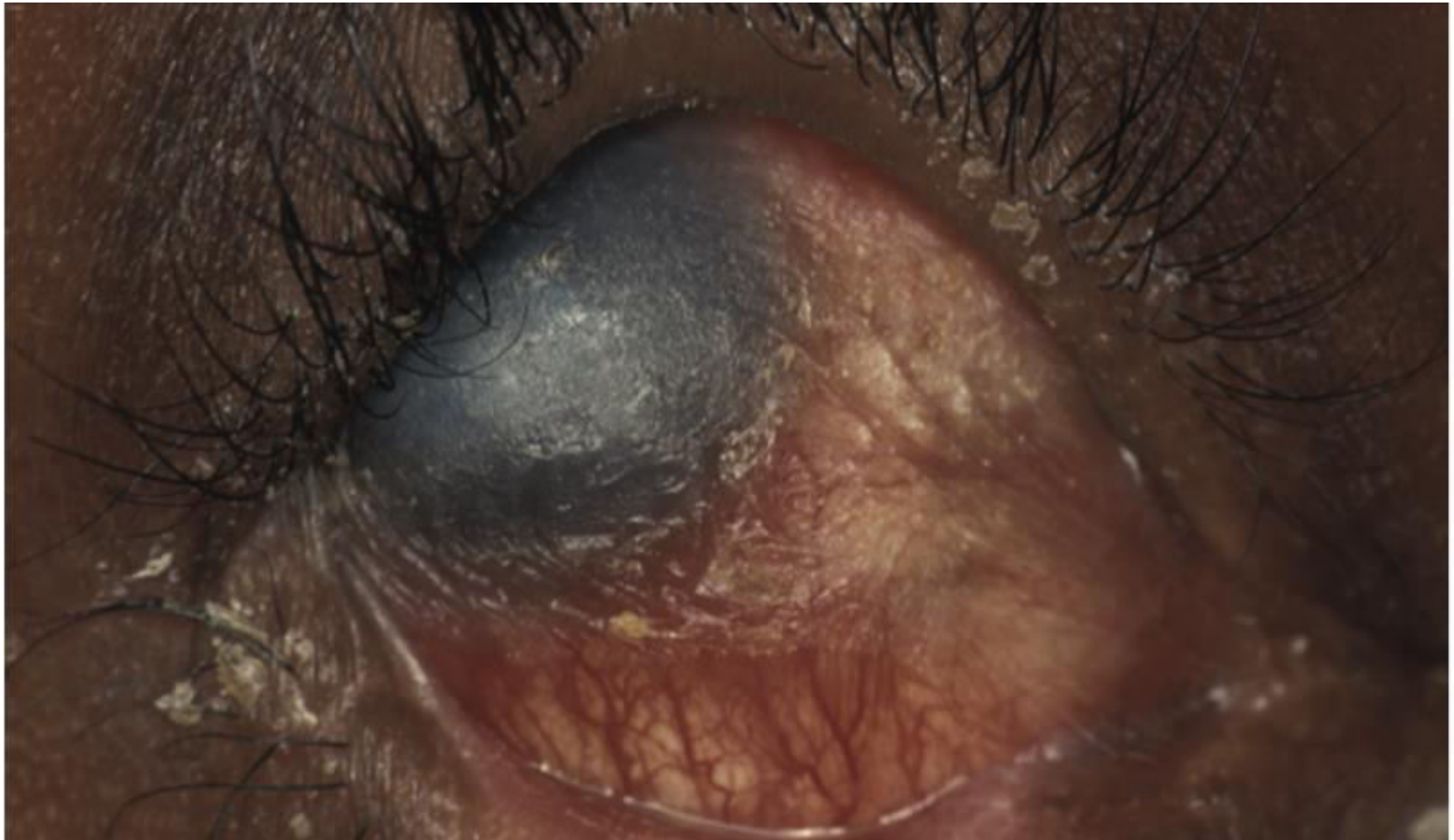


- Disruption of methionine metabolism
- Homocysteinemia milder clinical entity associated with cardiovascular and strokes
- Type 1 homocystinuria most prevalent inborn error of methionine metabolism (AR)
- Occurs 1 in 300,000 to 1 in 500,000
- Neurodevelopmental delay and failure to thrive
- Lens subluxates inferiorly or anteriorly
- High doses of vitamin B6 and cysteine supplementation

# Case 10 Steven Johnson



Moorfields  
Eye Hospital Dubai  
A BRANCH OF MOORFIELDS LONDON



07/05/2018

irfan.khan@moorfields.ae



# Steven Johnsons Syndrome



Moorfields  
Eye Hospital Dubai  
A BRANCH OF MOORFIELDS LONDON



07/05/2018

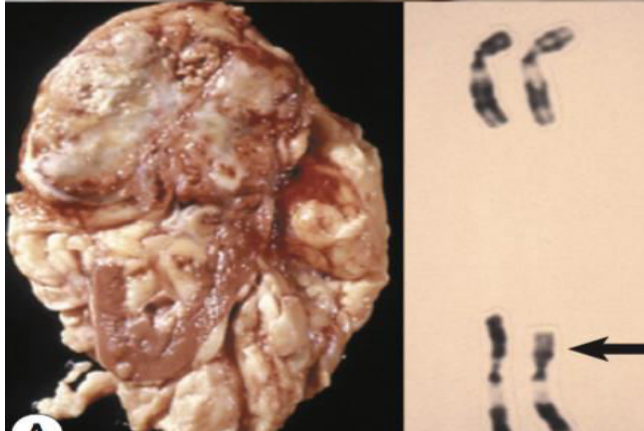
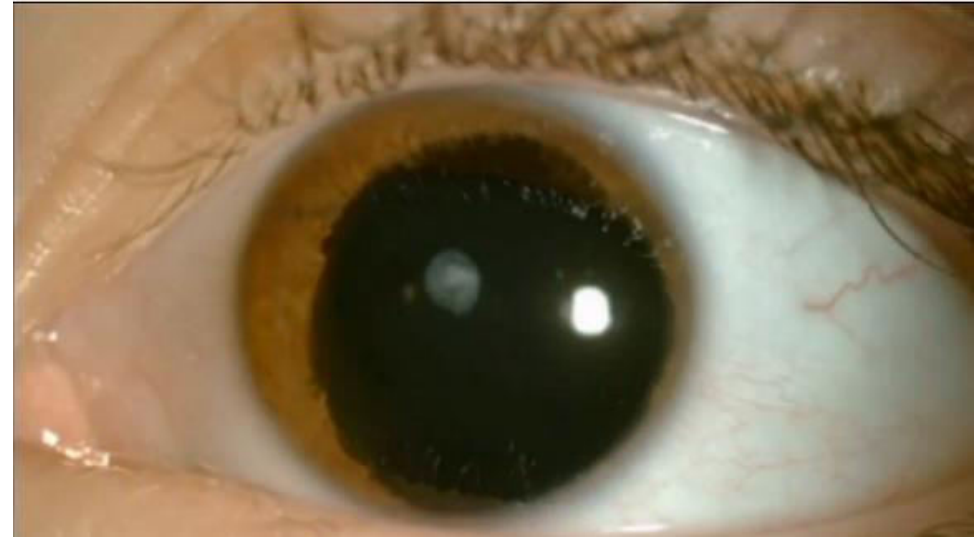
irfan.khan@moorfields.ae



# Case 11 Aniridia and Wilms Tumor



Moorfields  
Eye Hospital Dubai  
A BRANCH OF MOORFIELDS LONDON



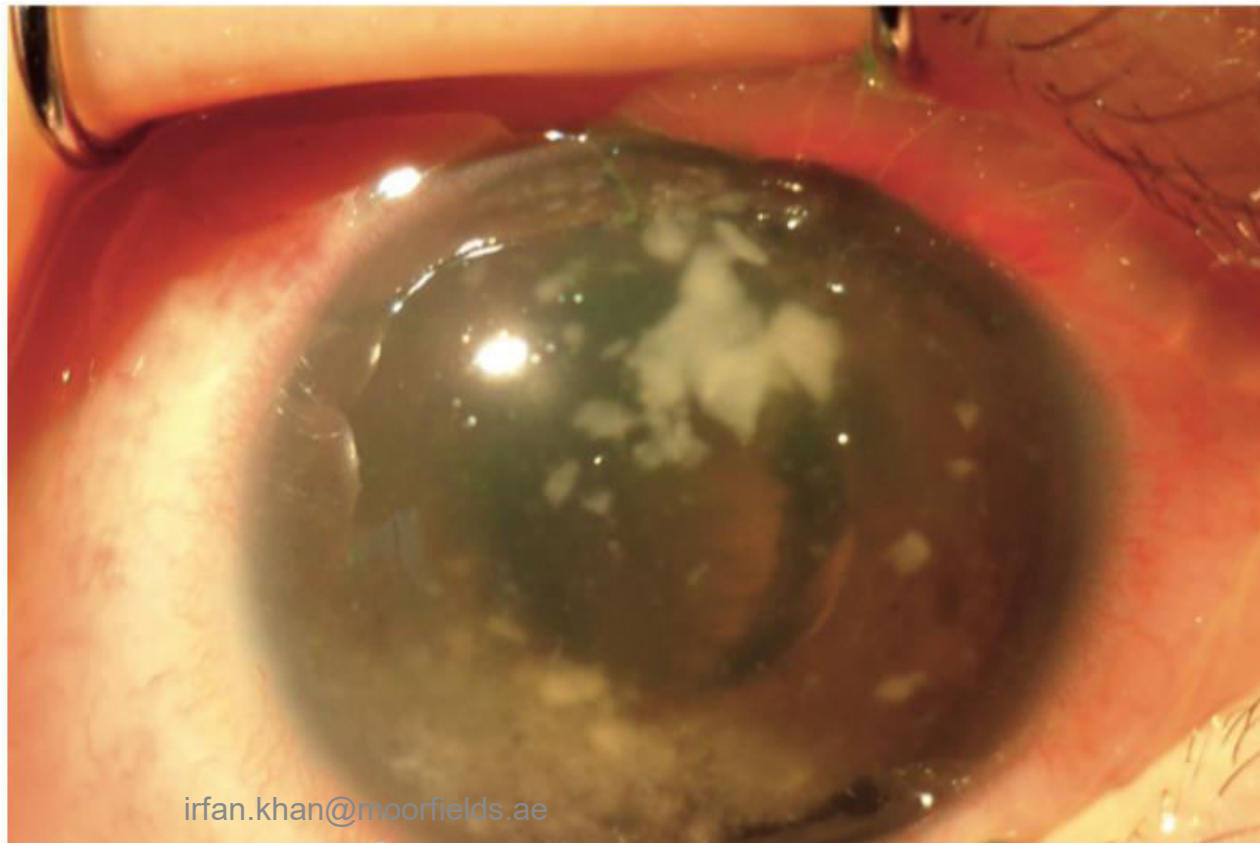
07/05/2018

irfan.khan@moorfields.ae

# Case 12 Retinoblastoma



Moorfields  
Eye Hospital Dubai  
A BRANCH OF MOORFIELDS LONDON



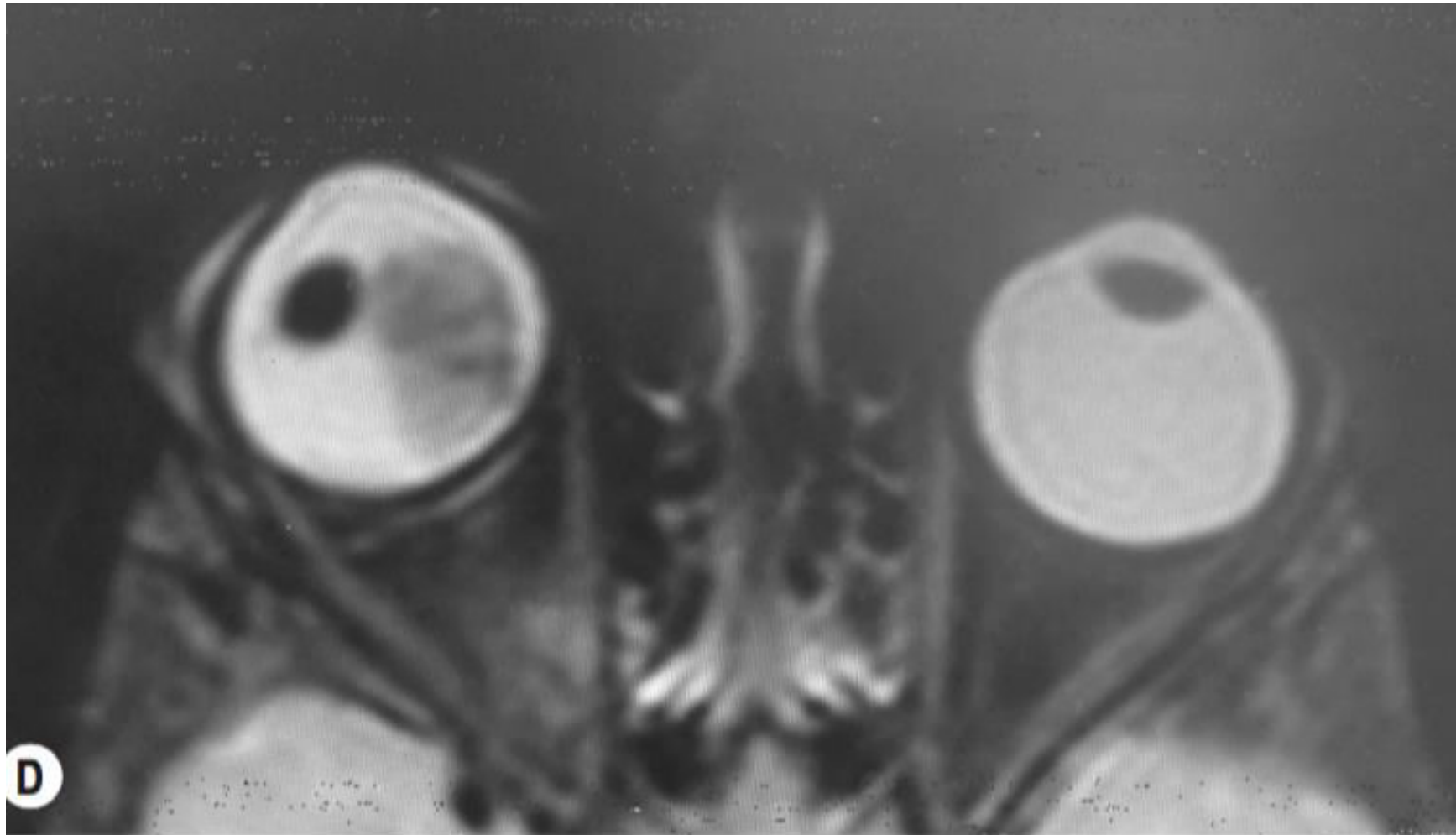
07/05/2018

irfan.khan@moorfields.ae

# Retinoblastoma MRI



Moorfields  
Eye Hospital Dubai  
A BRANCH OF MOORFIELDS LONDON



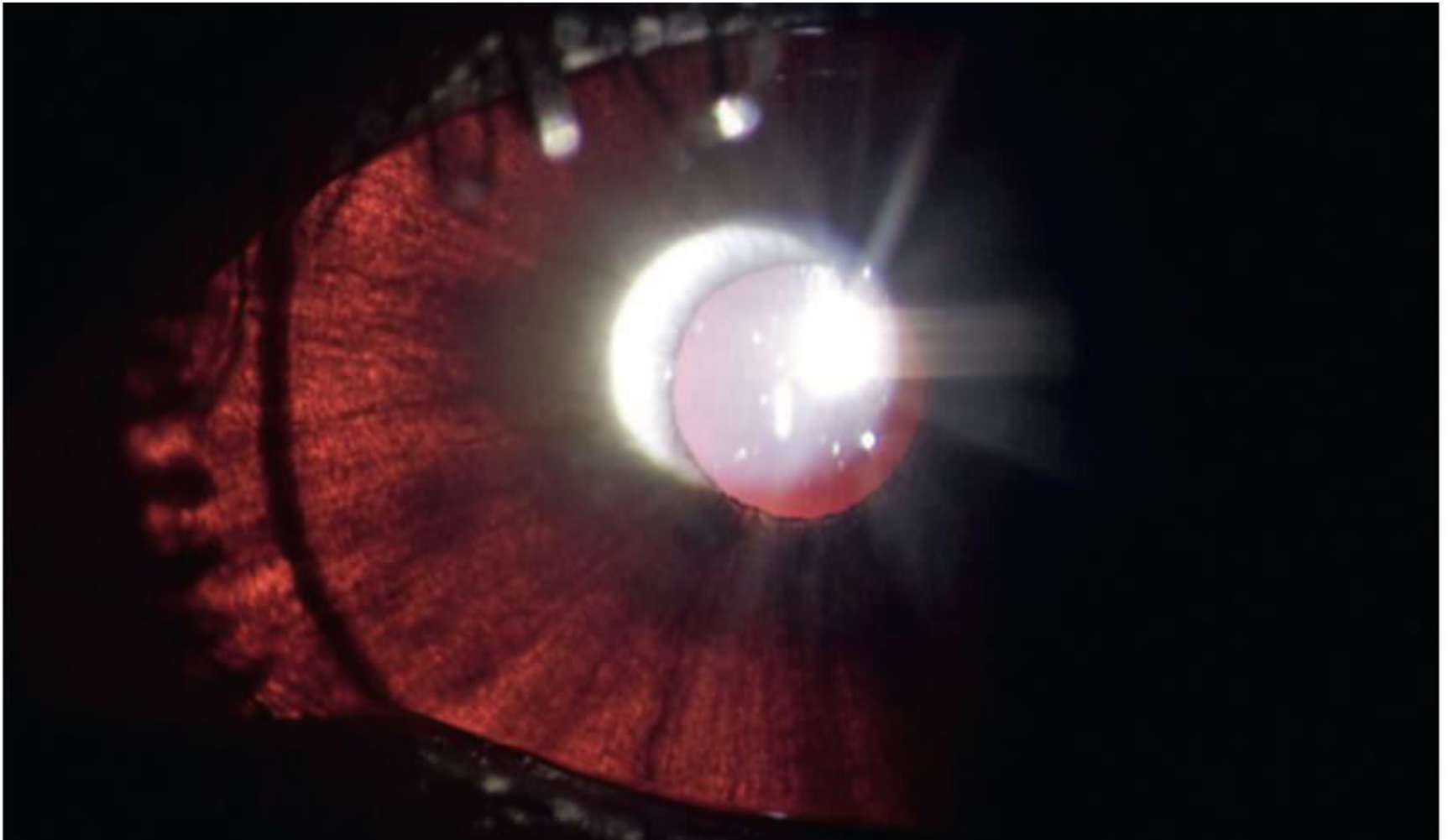
07/05/2018

irfan.khan@moorfields.ae

# Case 13 Albinism- Transillumination of Iris



Moorfields  
Eye Hospital Dubai  
A BRANCH OF MOORFIELDS LONDON



07/05/2018

irfan.khan@moorfields.ae





**(b) Albinism where the defect is not only of melanosomes but also of other related organelles (autosomal recessive inheritance and platelet storage pool deficit and bleeding tendency)**

Abbreviation	Long name	Gene product	locus	Phenotype distinctions
HPS1	Hermansky-Pudlak syndrome	HPS1	10q23	Commonest type of HPS, ceroid deposition lungs & gut, severe restrictive lung disease, founder effect in NW Puerto Rico
HPS2		ADTB3A	5q13	Neutropenia, frequent respiratory infections, impaired lymphocyte function, congenital dislocation of hips (dysplastic acetabulae), poor balance, conductive hearing loss, hemophagocytic lymphohistiocytosis
HPS3		HPS3	3q24	
HPS4		HPS4	22q11-q12	Ceroid deposition lungs & gut, mild to severe restrictive lung disease
HPS5		HPS5	11p15-p13	Hypercholesterolemia & hypertriglyceridemia
HPS6		HPS6	10q24	Respiratory and urinary tract infections, urinary and rectal incontinence, hearing loss, global developmental delay
HPS7		DTNBP1 dysbindin	6p22	Mild lung disease
HPS8		BLOS3	19q13.32	
CHS1	Chediak-Higashi syndrome	LYST	1q42.1-q42.2	Immunodeficiency, hemophagocytic lymphohistiocytosis, neurologic defects, hepatosplenomegaly

# Case 14 Left eye pushed Downwards



Moorfields  
Eye Hospital Dubai  
A BRANCH OF MOORFIELDS LONDON



07/05/2018

irfan.khan@moorfields.ae



# Post surgery for a Rhabdomyosarcoma



Moorfields  
Eye Hospital Dubai  
A BRANCH OF MOORFIELDS LONDON



07/05/2018

irfan.khan@moorfields.ae

# Case 15 Course feature Cloudy Cornea



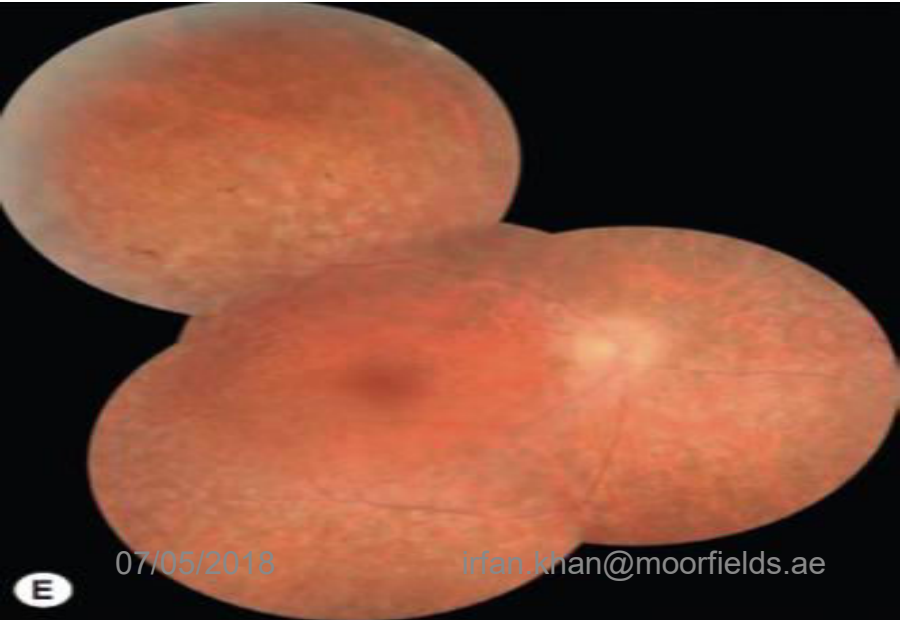
Moorfields  
Eye Hospital Dubai  
A BRANCH OF MOORFIELDS LONDON



07/05/2018

irfan.khan@moorfields.ae

# Case 16 Bardet Biedel Syndrome





- Obesity
- Polydactyly
- Hypogonadism
- Cognitive impairment
- Retinitis pigmentosa

Early onset before 3years

Legal blindness second decade of life

Rods and cones both affected



# Case 17 - Infantile Esotropia



Moorfields  
Eye Hospital Dubai  
A BRANCH OF MOORFIELDS LONDON



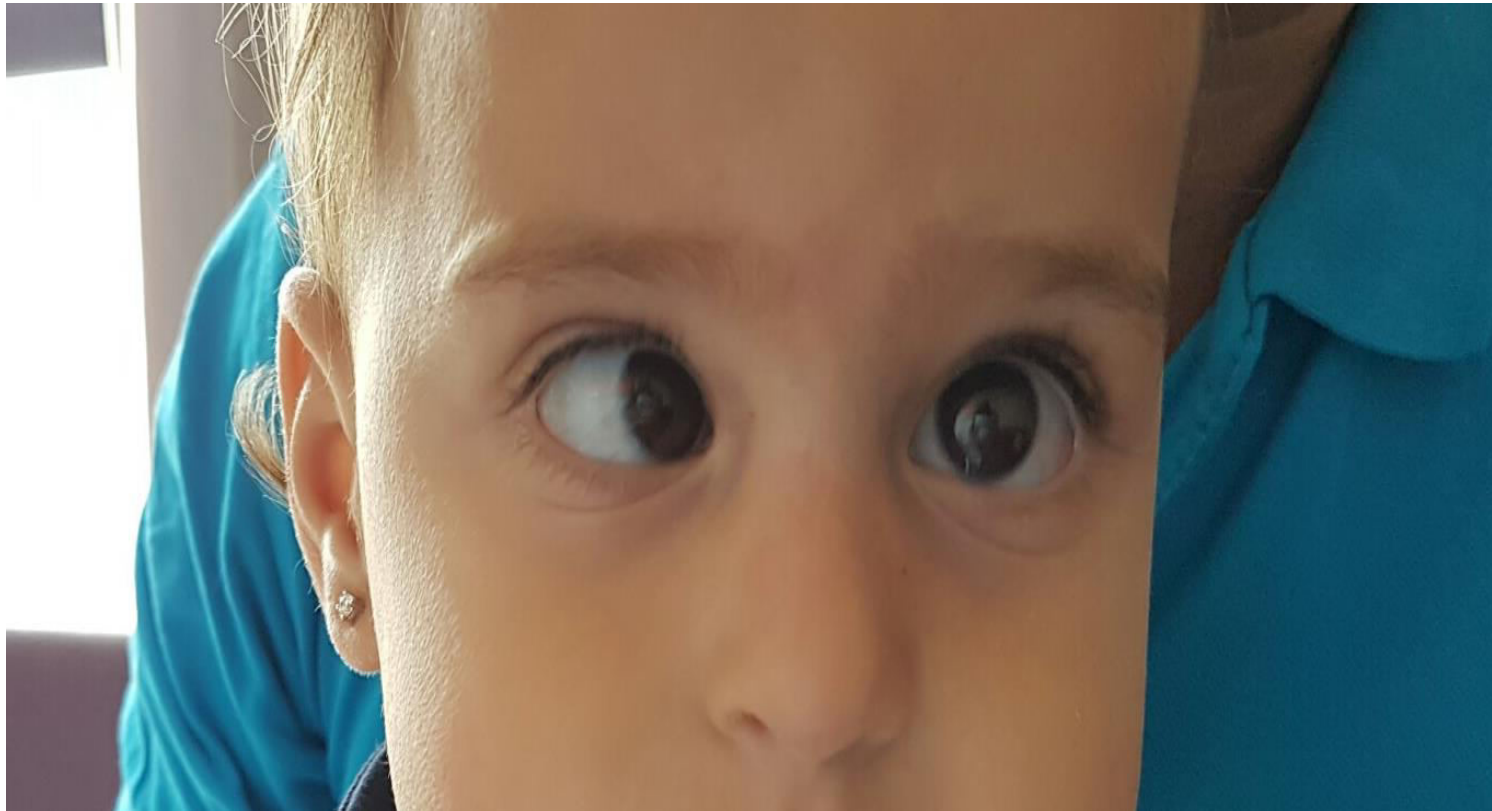
07/05/2018

irfan.khan@moorfields.ae

# Infantile Esotropia



Moorfields  
Eye Hospital Dubai  
A BRANCH OF MOORFIELDS LONDON



07/05/2018

irfan.khan@moorfields.ae





# Day 1 Post Op



Moorfields  
Eye Hospital Dubai  
A BRANCH OF MOORFIELDS LONDON



07/05/2018

irfan.khan@moorfields.ae

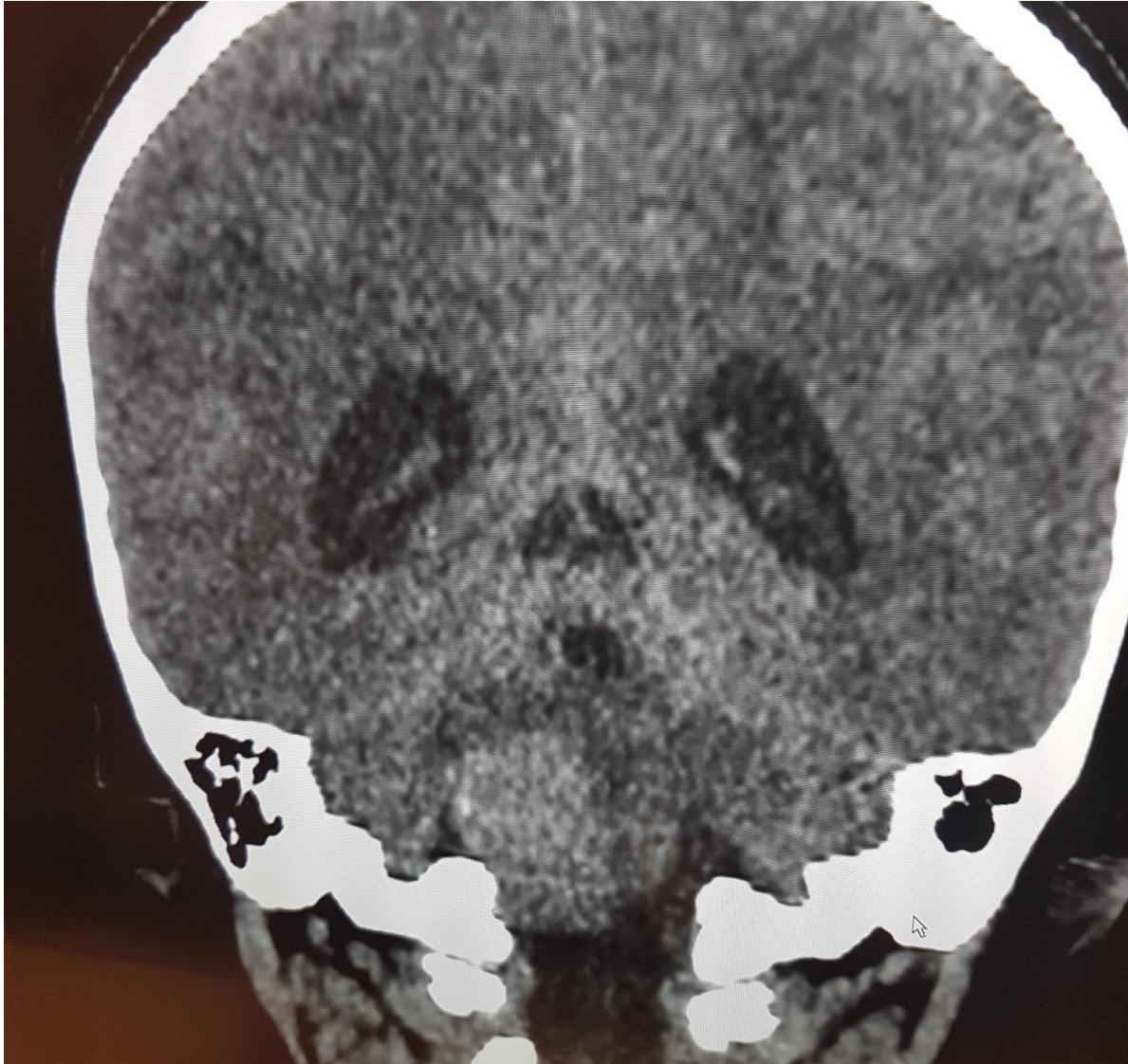


- 3.5 yrs old
- JIA on methotrexate
- Four weeks onset esotropia
- Seen by local ophthalmologist X 4
- Given hyperopic glasses
- Reassured
- Parents anxious came in for a 5<sup>th</sup> opinion

# Image 1



Moorfields  
Eye Hospital Dubai  
A BRANCH OF MOORFIELDS LONDON

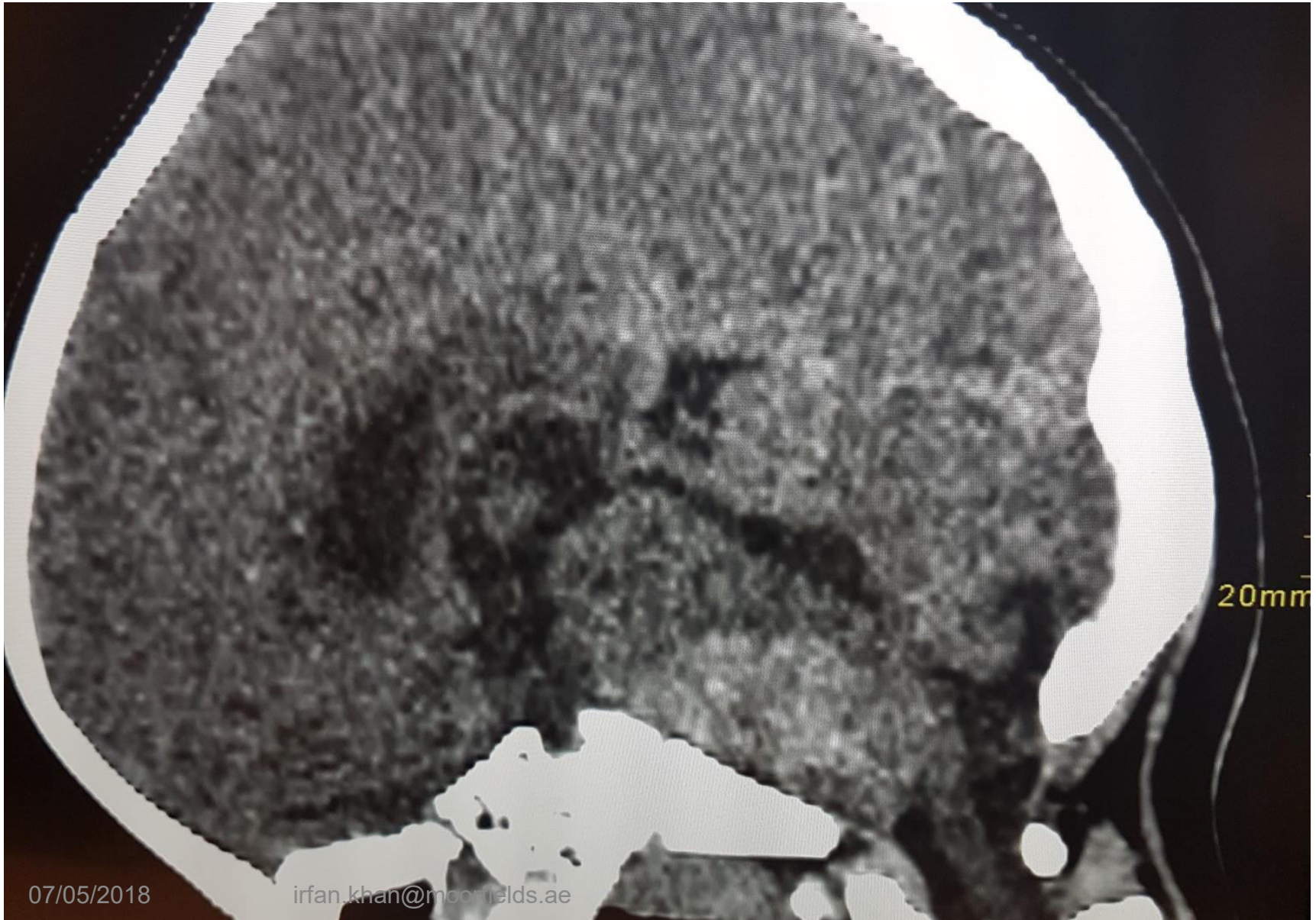


07/05/2018

[irfan.khan@moorfields.ae](mailto:irfan.khan@moorfields.ae)



# Image 2



07/05/2018

irfan.khan@moorfields.ae

# Case 19



Moorfields  
Eye Hospital Dubai  
A BRANCH OF MOORFIELDS LONDON



07/05/2018

irfan.khan@moorfields.ae



# Case 20



Moorfields  
Eye Hospital Dubai  
A BRANCH OF MOORFIELDS LONDON



07/05/2018

irfan.khan@moorfields.ae

# Case 21



Moorfields  
Eye Hospital Dubai  
A BRANCH OF MOORFIELDS LONDON



07/05/2018

[irfan.khan@moorfields.ae](mailto:irfan.khan@moorfields.ae)

# Congenital Glaucoma



Moorfields  
Eye Hospital Dubai  
A BRANCH OF MOORFIELDS LONDON



07/05/2018

irfan.khan@moorfields.ae

# Case 22 Capillary Haemangioma with Right Ptosis



Moorfields  
Eye Hospital Dubai  
A BRANCH OF MOORFIELDS LONDON



07/05/2018

irfan.khan@moorfields.ae



# After Oral Propranolol



Moorfields  
Eye Hospital Dubai

MOORFIELDS LONDON



07/05/2018

irfan.khan@moorfields.ae

# Summary



Moorfields  
Eye Hospital Dubai  
A BRANCH OF MOORFIELDS LONDON

- Take your time
- History
- Second opinion
- Knowing it all !
- Counseling parents





Thank You

[Irfan.khan@moorfields.ae](mailto:Irfan.khan@moorfields.ae)

Moorfields Eye Hospital