CASE FILES: THE RETINA CHRONICLES

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Charles

- BCVA 20/20 with hyperopic RX
- Entrance tests unremarkable
- Anterior seg: wnl
- Posterior seg:

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Retinoschisis

- Defined as splitting of the neurosensory retinal Layers
  - Typically in outer plexiform layer
- Variability: large, small, bullous, flat, bilateral, unilateral, progressive ,non-progressive
- Two major types
  - Acquired
  - x-linked (XLRS)

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Acquired retinoschisis

- Usually benign and non-progressive
  - Myopic pts tend to be more progressive than hyperopic pts
- Asymptomatic, found on routine DFE, but may cause VF defect
- Incidence:
  - 3.9% in pts 60-80
- Most commonly affects inferotemporal retina
- Bilateral 33-82% of time

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Schisis vs RD

- Schisis:
  - More translucent with visible vasculature
  - Less flexible
  - Well demarcated borders
  - Overall smoother appearance
  - Should have absolute Vf defect vs relative with RD
- B scan/OCT can be helpful
  - OCT often difficult to image due to location

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Retinoschisis

- Can have outer or inner wall breaks
  - Outer:
    - larger, often have ring of pigment
    - 11-24% of time
  - Inner:
    - smaller
    - Look like atrophic holes
  - Either associated with increased risk for detachment, so retinal consultation advised
  - Inner and outer together very dangerous
- if no holes, generally benign and can be monitored

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Retinoschisis

- Very rare to have detachment into macula area
- Prophylactic Laser treatment has not been shown effective in most studies to halt progression
- Cataract surgery and pvd do not seem to have adverse effect
- If progresses to detachment, retinal surgery indicated
  - Only about 0.05% to 2.2% of casers
  - Typically respond poorly to surgery
- Most are benign and can be monitored yearly unless holes, enlargement, or symptoms

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Idiopathic Central Serous Chorioretinopathy (ICSC)

- Condition of unknown etiology
- Anxious males: 3:1 to 10:1
  - Incidence doubles in women 30-40 vs 20-30 y/o
- History of emotional stress
- "Type A personality"
- Common Caucasians, Hispanics, Asians

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ICSC

Clinical Features

- Detachment of sensory retina
  - Due to leakage from small underlying PED
- Absence of foveal reflex
- Vision rarely less than 20/25
- Patients often report micropsia

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Central Serous Chorioretinopathy (CSR)

2 Main Types

- Common classic CSC
- More widespread alteration of the RPE with chronic shallow SRF
  - Chronic CSC
  - May be associated with chronic corticosteroid use

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Corticosteroids and CSR

- Strong relationship with increased cortisol levels
  - Steroid users
  - Organ transplant
  - Medical conditions requiring steroid: SLE, RA
- Pregnant women
  - Increased levels of free circulating endrogenous cortisol

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Case

- **CC: 56 year old African American female presents for a retinal evaluation of AMD from her Optometrist. The patient complains that she has trouble seeing darker faces. In addition, she and her husband often argue over the color of an item. Patient states that she feels she drives fine during the day, but has to put on glasses at night because she has more difficulty seeing in the dark. Patient has a history of ocular migraines, but does not get them frequently. Also, the patient states that she was punched in her right eye three years ago.**
- **Medical History: Hypothyroid and Systemic Hypertension and takes Levothyroxine and Hydrochlorothiazide orally. No eye drops.**
- **Best corrected VA: 20/20-3 OD and 20/40-2 OS at distance**
  J1-1 OD and J10 OS at near
- **IOP: 14 mmHg OD and 15 mmHg OS using Goldman Applanation Tonometry**
- **No previous ocular surgeries or treatments have been performed on the patient.**
- **Slit lamp exam was remarkable for 1+NS in both eyes, but all other anterior segment structures were normal.**
- **Posterior segment exam shows clear vitreous, C/D 0.2 OD and OS, normal vessels and SDE peripheral exam.**
  - **See next slide for macular scans and findings**

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What is Taxol?

- Taxol, a taxane, a class of antimicrotubule agents and chemo drugs used to treat cancer. Paclitaxel is the active ingredient which was originally isolated from the bark of the Pacific yew tree in 1971.
- Used to treat breast CA (HER2-positive)-effective against early and advanced stage BCA. Also effective against advanced ovarian, lung, bladder, esophagus, and head and neck CA.
- Given Intravenously every 1-3 weeks and is often combined with other chemotherapy drugs.
- Many side effects due to disruption of microtubule function: peripheral neuropathy, arthralgias, myalgias, liver and retinal damage

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Better safe than sorry

- Genetic testing was ordered to rule out any chance of patient having Age Related Macular Degeneration or any other Inherited Retinal Disease
- And the test results were negative for any inherited retinal disease findings

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Purtscher retinopathy

- 1st described in 1910 by otmar purtshcer in a man who fell from a tree and suffered cranial trauma and retinal findings
- Retinal Findings:
  - Cotton wool spots (93%)
  - Retinal hemorrhages (65%)
  - Purtscher flecken (63%)
    - Pathognomonic finding
    - Polygonal areas of retinal whitening with a clear demarcating line between the affected retina and normal retinal vessels

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Purtscher retinopathy

- Characterized by painless decreased VA
- May appear at same time of event or 24-48 hrs after
- Bilateral 60%
  - virtually always bilateral if associated with acute pancreatitis
- Typically associated with central, paracentral , or arcuate scotomas on VF

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Purtscher retinopathy

- Classically associated with compression trauma
- Long bone fracture
- **Acute pancreatitis**
- Renal failure
- Preeclampsia and childbirth
- Barotrauma
- Retrobulbar anesthesia
- Valsalva
- Shaken baby syndrome
- And more

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Purtscher retinopathy

- Differential includes
  - Branch retinal artery occlusion
  - Central retinal artery occlusion
  - Commotio Retinae

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Treatment

- Observation
- Treatment of underlying condition, ie pancreatitis
- Intravenous, high dose steroids: no evidence
- Papaverine hydrochloride: no evidence
- Hyperbaric oxygen: no evidence

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Treatment

- 40% resolve within 2 mos with no sequelae
- Potential sequalae:
  - Optic atrophy 64%
  - Macula mottling 23%
  - Retinal thinning 14%
  - Narrowing of retinal arteries 4%
- Visual recovery is variable

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Acute pancreatitis: diagnosis

- Sudden inflammation of the pancreas
- Abdominal pain
- Lipase or amylin 3x upper limit of normal
  - Digestive enzymes in the pancreas
  - Lipase: Normal Range: 11-82
  - Our pt: 1721 u/l (H)
- Abnormal abdominal ultrasound (MRI or CT)

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Acute pancreatitis: common Causes

- Gallstones
- Auto-immune disease
- Damage from surgery or injury
- Cystic fibrosis
- Kawasaki disease
- Viral infection (ex mumps)
- Reye's Syndrome
- Certain medications or drugs
- **ALCOHOL ABUSE (30% of all cases)**

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Acute pancreatitis: treatment

- May need hospital admittance for acute phase with pain relivers and Iv Fluids
  - Antibiotics if secondary infection
- Mild to moderate pancreatitis self limiting within 1 week
  - Severe cases several weeks
- **Avoid heavy alcohol use**
  - **Anyone who has had one episode of acute pancreatitis should stop drinking entirely**

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Valsalva Retinopathy

Valsalva occurs when a person tries to **exhale air forcibly** with a **closed glottis** (windpipe) so that no air goes out through the mouth or nose

Sudden increase in intrathoracic or intra-abdominal pressure occurs as a result of this forced exhale

Hemorrhage occurs due to spontaneous rupture of superficial retinal capillaries

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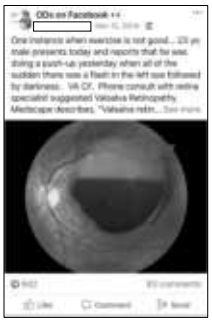
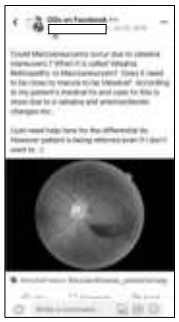

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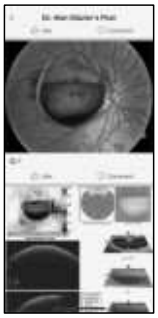


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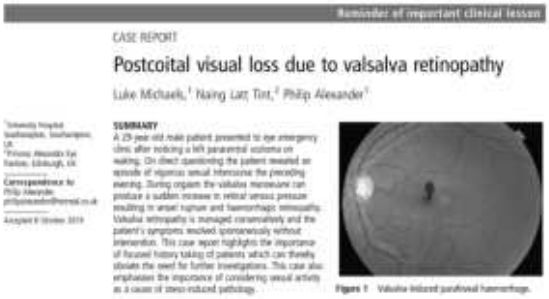
Reminder of important clinical lesson

CASE REPORT

Postcoital visual loss due to valsalva retinopathy

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SUMMARY
A 25-year-old male patient presented to eye emergency clinic after noticing a left paracentral scotoma on waking. On direct questioning the patient revealed an episode of vigorous sexual intercourse the preceding evening. During orgasm the valsalva manoeuvre can produce a sudden increase in retinal venous pressure resulting in arteriolar rupture and haemorrhagic retinopathy. Valsalva retinopathy is managed conservatively and the patient's symptoms resolved spontaneously without intervention. This case report highlights the importance of focused history taking of patients which can thereby obviate the need for further investigations. This case also emphasises the importance of considering sexual activity as a cause of stress-induced pathology.

Figure 1 Valsalva-induced parafoveal haemorrhage.

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Case Report

Valsalva Retinopathy Associated with Sexual Activity


Figure 1: Massive subinternal limiting membrane (ILM) haemorrhage secondary to Valsalva manoeuvre

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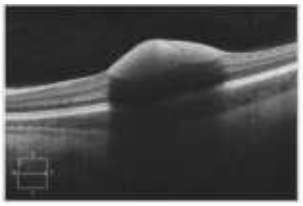
Where is the hemorrhage?

- Pre-retinal
- Subhyloid
- Sub – ILM



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Sex-Induced Valsalva Retinopathy

Learning points

- Focused history taking is of great diagnostic importance because with certain topics, patients will not necessarily volunteer the history.
- If a suspicion exists that patient presentation is linked to sexual activity, physicians should be aware of the sensitivities of taking a sexual history and be confident in their ability to do so.
- Valsalva retinopathy can be caused by any physical stress, including sexual activity.
- Sex-induced valsalva retinopathy has been reported and is thought to be rare but the true incidence is unknown.
- Valsalva retinopathy is a self-resolving condition with an excellent prognosis.

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Valsalva Retinopathy: Treatment

- Spontaneous resolution – slow absorption of the hemorrhage
- YAG laser lysis
- Pars Plana Vitrectomy (PPV)

- He returned for follow up ~ 1 week later with no improvement
  - Then was lost to follow up

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The Moral of the Story…

- Sometimes its not so important to know why

- Remember –

A Closed Mouth Gathers No Feet

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Valsalva Retinopathy

- Valsalva occurs when a person tries to **exhale air forcibly** with a **closed glottis** (windpipe) so that no air goes out through the mouth or nose
- Sudden increase in intrathoracic or intra-abdominal pressure occurs as a result of this forced exhale
- Valsalva maneuver occurs from various day-to-day activities that cause straining such as coughing, sneezing, vomiting, exercise, blowing on musical instruments, among others[1].

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Case :

- **CC: On May 18, 2011 a 71 year old Caucasian male presented from his Optometrist's office. The Optometrist was concerned that there may be a problem with the retina due to not being able to correct the patient fully with glasses in either eye. The patient was frustrated because he could not pass the vision part of his driver's license testing.**
- **Best corrected VA: 20/30+1 OD and 20/70+1 OS at distance**
  20/50 OD and 20/400 OS at near
- **IOP: 17 mmHg OD and 13 mmHg OS using Goldman Applanation Tonometry**
- **Past Ocular Surgery: Phaco/PCIOL OD**
- **Slit lamp exam was remarkable for PCIOL OD and 3+NS OS, but all other anterior segment structures were normal.**
- **Dilated Fundus Exam show PVD OD and clear vitreous OS, C/D 0.35 OD and 0.40 OS, normal vessels and SDE peripheral exam revealed reticular pigmentary changes in both eyes.**
  - **See next slide for macular scans and findings**

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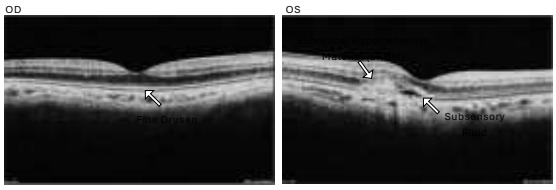
OCT at initial visit on 5/18/2011



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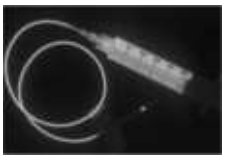
What about Fundus Autofluorescence or Fluorescein Angiography?

- In 2011, FAF was not widely used for retinal exams.
- The Fluorescein Angiography was attempted, but the patient began to feel light headed, broke out in a rash and vomited-images were not captured as patient was treated for severe reaction to FA dye.

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Diagnosis and plan of treatment:

- Early Dry AMD in the right eye and Wet AMD with active CNVM in the left eye
- Treatment plan included:
  - IV Bevacizumab (IVA)
  - q 6 weeks
  - 4 treatments
  - Then re-evaluate

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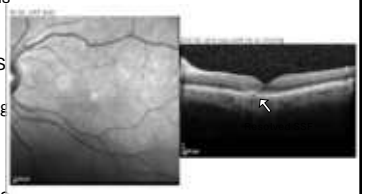
Follow up exam 11/11/2011

- CC: Vision seems to be improving, but patient complains of "granules in left eye".
- Dist BCVA: 20/25 OD, 20/40 OS
  Near BCVA: J2 OD, J5 OS
- IOP: OD 15mmHg, OS 14 mmHg
- SLE: PCIOL OU, 1+PEE OS
- DFE: Stable, except controlled CNVM
- Plan: Continue IVA q 6 weeks OS and PF AT QID OU for dry eye



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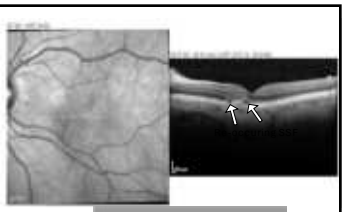
8/21/2012

- A total of 10 IVA injections were administered with good CNVM control in the left eye.
- At this time VA remained 20/30 in the left eye, however a sliver of SSF remained that showed the resilience of the net of vessels.
- The drug was then switched to IV Aflibercept (IVE) on 8/21/2012 for the persistent CNVM in the left eye.

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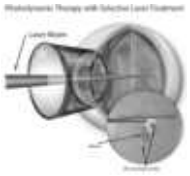
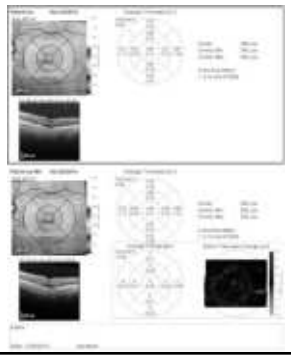
11/19/2013

- **The IVE x 11 was given q 6-8 weeks in the left eye.**
- **BCVA OD: 20/20, OS 20/50**
- **Persistent SSF is noted on the OCT OS.**
- **Plan: Stat Photo Dynamic Therapy was employed in the left eye along with continuing IV Eylea.**

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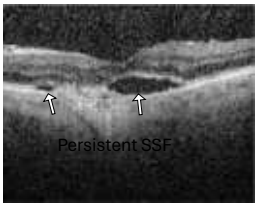
62!!!! More IVE injections in the left eye

- A total of 62 more IV Eylea injections were given between 11/19/2013 and 12/17/19 OS.
- On 12/17/2019, the BCVA was found to be:
  OD 20/25 (Dry AMD)
  OS 20/60 (persistent CNVM)



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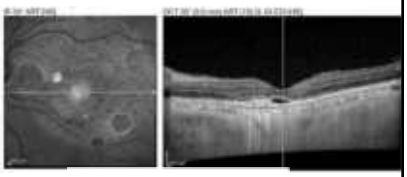
Change of plan…

- IV Brolucizumab-dbll (IVB) then injected on 12/17/2019 -7/28/2020 q 6-8 weeks for a total of 5 IVB in OS.
- VA remained stable at this point.

  BCVA OD: 20/25+1

  OS: 20/50-2



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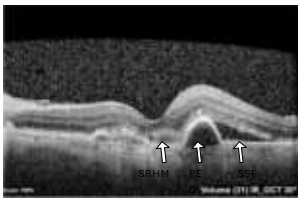
Back to Eylea, but now in a Pre-Filled Syringe

- On September 21, 2020 IVE PFS was given in the left eye for a total of 12 injections OS given q6-8 weeks with the last given 5/23/22.
- BCVA remained relatively stable at 20/30+2 OD, but worsened to 20/400 OS.
- The OCT reveals worsening CNVM OS.
- Why Pre-Filled Syringe??



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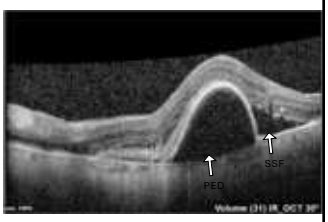
Yet another type of med given on 6/28/22

- IV Faricimab-svoa (IVV) was given on 6/28/22 in the left eye.
- Vision in the left eye has now gone down to CF@4 feet.
- The CNVM is worsening in the left eye.
- The right eye has remained dry AMD with stable VA.



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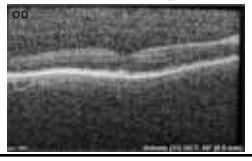
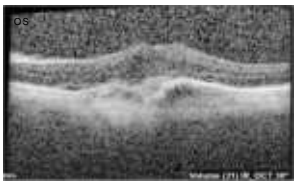
What the heck, let's try Susvimo…

- **Finally, the Susvimo implant was placed and filled on 8/1/22.**
- **The patient has been able to have successful refills q 6 mos through 7/30/24 for a total of 5 treatments in the left eye.**
- **Vision in the left eye is 20/200 as of 7/30/2024.**
- **The right eye has remained dry with no treatment needs and VA of 20/40.**





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What is Susvimo?

- Susvimo is a Port Delivery System with ranibizumab.
- SUSVIMO® is the first and only treatment for nAMD that continuously suppresses VEGF in the eye for 6 months
- Refillable implant
- Only 2 treatments per year

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Who qualifies for Susvimo?

- SUSVIMO (ranibizumab injection) is indicated for the treatment of patients with neovascular (wet) age-related macular degeneration (AMD) who have previously responded to at least 2 intravitreal injections of a vascular endothelial growth factor (VEGF) inhibitor medication.

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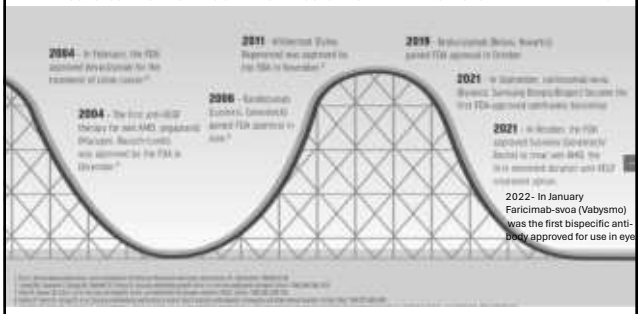
THANK YOU FOR JOINING ME ON THIS CRAZY ROLLER COASTER OF TREATMENT OPTIONS…THE RIDE NEVER ENDS!!



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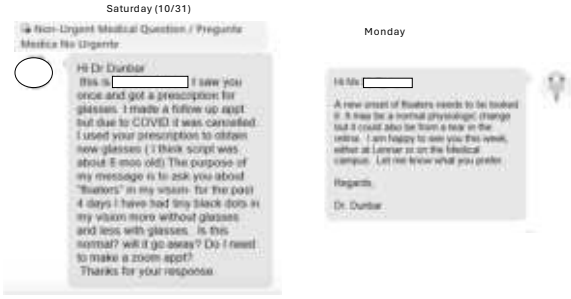

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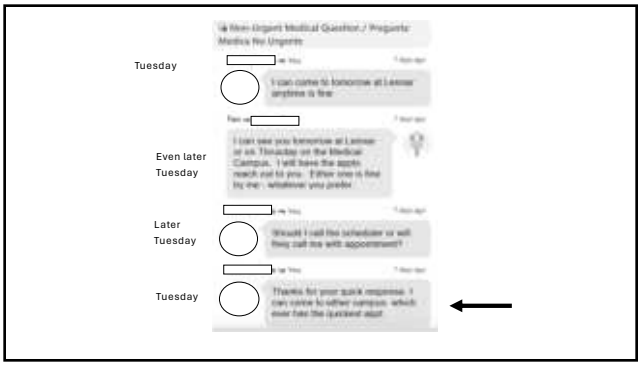

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Predictive Factors for Poor Visual Outcome

- Initial presenting visual acuity – worse visual acuity
- Duration of the macular detachment
  - Longer the mac-off RD -> worse the visual outcome
- Height of the retinal detachment

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Timing of **Macula-Off RD** Surgery

- Best visual outcomes are when the surgery is performed within **7 days** of the macular detachment
- Meta-analysis have shown that a delay of **more than 3 days** was associated with statistically worse final visual outcome
  - Eyes that had surgery within 3 days averaged a final visual acuity of around 20/30
  - Eyes that were operated between 4-7 days averaged a final visual acuity of around 20/70.

AJO 2022: 244; 19-20

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