VA Grand Rounds Course # 40042

Instructor:

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Section:

Systemic Disease

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Qualified Credits:

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COURSE DESCRIPTION:

This course highlights challenging presentations of ocular disease that present in a high volume ocular disease VA practice. The goal is to learn diagnosis and management strategies of common and uncommon ocular diseases.

LEARNING OBJECTIVES:

- Understand diagnosis and management of CRAO
- Understand difference between optic nerve pallor and excavation
- Discuss the role for OCT in tractional macular disease
- Learn management strategies for idiopathic intracranial hypertension

Course begins on page 2
Hello, and welcome to Pacific University Online Continuing Education. My name is Anthony DeWilde, and I’m an optometrist at the Kansas City VA Medical Center. What I’m going to do for the next hour is walk you through a Grand Rounds presentation of some interesting patients I’ve seen at the VA. Some of these patients are probably similar to what you’ve seen on your own, and some of them are probably quite different. My hope in this grand rounds presentation is to take it above and beyond certain grand rounds presentations. I feel like sometimes there’s this tendency to stump the people in the audience, and I’m not really as interested in that as much as I am interested in highlighting my thought process through diagnosis, through treatment and referral. Hopefully you can pick up some ideas of how to treat certain eye diseases, or thinking broadly about how to diagnose. Let’s get started here.

**Patient 1**

My first patient is a 72 year old African American male who comes in with a complaint of blur for 3 months. He wasn’t brand new to us – we had seen him 10 years prior, but we hadn’t seen him since that exam, which is troublesome, because this patient has mixed-mechanism glaucoma. He had an LPI in both eyes around 10 years ago. Historically, he had been taking Xalatan qhs OU, but has not been taking it since he was lost to follow-up, because we hadn’t renewed it. In addition, he has a history of blunt trauma to his right eye.

His medical history, as you can see in Table 1, indicates that he is not the healthiest patient out there. He has some kidney disease, a stroke twice, hypertension and hyperlipidemia. Not terribly different from some of the 70 year old patients who are seen at the VA, but he does have a lot of things going on. His medications include Amlodipine, Atenolol, HCTZ, Simvastatin, and ASA, which reflect his complicated medical history of stroke and high blood pressure.

When I saw him, his blurred vision OD was 20/320 with the best glasses I could find him. His left eye was pretty good, at 20/25. We noted a right APD. His anterior segment was normal, and he just had a mild cataract OU – definitely not bad enough to account for 20/320. Before I proceed, 20/320 is measured with ETDRS chart or Lighthouse Chart – there is really no 320 at Snellen chart. At 10 feet, he was reading the 160 line, thus 20/320. The Snellen would be 20/400.

We did gonioscopy, just to look and see if everything was OK. He had some old PAS there. The angle was narrow, but he had an LPI to help resolve that narrow angle. His pressure the day I saw him, untreated, was 14 in each eye.

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<tr>
<th>Medical History</th>
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<tr>
<td>Hypertension</td>
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<td>Cerebrovascular Accident x2</td>
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<td>Hyperlipidemia</td>
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<td>Kidney Disease</td>
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Table 1

The posterior segment is where things start to get interesting. I looked at his right eye, which is the worse of the two eyes, and I noted that not only is that the smaller of the two nerves, but it’s pallid. If this was from glaucoma, we would expect the 0.9 nerve to be the one with the vision loss, right? Maybe he’s got an inferior notch or excavated enough nerves that he has central vision/fixation loss? But that wasn’t the case – it was his right eye that was worse, and his nerve was pallid.

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<th>Posterior Segment</th>
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<tr>
<td>Optic nerve:</td>
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<tr>
<td>OD: 0.75 – Pallor</td>
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<tr>
<td>OS: 0.90</td>
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<tr>
<td>No Maculopathy</td>
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<tr>
<td>No Vasculopathy</td>
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<tr>
<td>Peripheral Retina Normal</td>
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Table 2
We need to resolve that in our mind. Why would the smaller-cup nerve be worse? Why would the smaller nerve be pallid? He had no maculopathy, no vasculopathy, nothing else that would explain this vision loss in the right eye.

Let’s take a brief moment to talk about pallor versus excavation. Pallor is not something that you are going to see from glaucoma, except in one specific instance, and that’s if the patient has a history of very high IOP – we’re talking 50’s, 60’s and beyond. What the thought process is with this patients is that the pressure got so high that they had a central retinal artery occlusion (CRAO). Maybe a profound CRAO or maybe a mild one, but it can leave their nerve looking pallid. Figure 1 is a stereo photograph of a patient with a pallid nerve. When they first look at this nerve, a lot of people think “Oh, that’s glaucoma.” In fact, in this patient specifically, it fooled us for a while, too.

This patient has a large cup-to-disc ratio, as you can see. Someone might grade that as a 0/6 or above. If you look at the rim, specifically, which is where we’re looking for pallor, that’s where you can see that this nerve has a little bit of a discoloration – it’s white-ish in color. In a nerve that is glaucomatous, we are expecting excavation beyond pallor, but this nerve actually has a large cup-to-disc ratio, and I might say that it could be a naturally large cup-to-disc ratio, but the rim tissue that is remaining is pallid. Sometimes in glaucoma you can get something called ‘pseudo-pallor’, and what happens with that is that when the nerve tissue, the rim tissue, excavates away, that leaves more cup left over, and naturally the cup is looking at empty space, lamina, which is much whiter. Thus, it can be pseudo-pallid because you’re looking at more lamina, and not rim, thus it appears pallid. Thus, we need to make sure we are defining pallor correctly. Pallor is when the remaining rim that is there isn’t healthy; is pallid and white. Not necessarily when the cup is white.

Figure 2 shows the other eye of this patient. You can see it’s naturally a large cup-to-disc ratio in this eye, as well. It may be glaucomatous – the cup is definitely enlarged, but look at the color of the rim on this nerve. This rim looks quite healthy – it has normal pink/yellow-ish hue to it, where the nerve in Figure 1 has a
white-ish sort of sick look to it. We should not confuse pallor an excavation – these are two different things.

Let me give you another example. Look at the temporal rim of this patient in Figure 3 [Top] compared to the nasal rim of this patient. The nasal rim looks pretty normal, whereas the temporal rim is quite pallid. It looks much sicker.

Let’s compare it to the companion eye. (Figure 3 [Bottom]) This eye, as you can see, has a beautiful nerve fiber layer (NFL), it’s got a very healthy looking nerve; nothing about it looks sick or unhealthy.

Looking back at Figure 3 [Top], this one might have a larger CD ratio, but when you look at the rim that is remaining there, that is actually pallid rim, specifically temporally, with a little bit superior and inferior. Just from this regular photo that is not red-free, altered, or anything like that, the NFL even here looks a little bit sicker.

Figure 4 [Top] shows us another temporal rim that is very pallid. You can see the lamina here, and you can see that there is rim tissue that looks reasonable – it’s not great, but it’s reasonable. Specifically looking temporally, that is quite pallid. Figure 4 [Bottom] shows us the other eye of the same patient, and what a contrast, right? Really healthy NFL, really healthy rim tissue. It’s a larger cup, and the cup looks pale – it always does – but the remaining rim is not.

The differential on our patient here was glaucoma. Again, as I mentioned earlier, I had my misgivings about that, because if we were expecting glaucoma, we would anticipate the worse vision being in the eye with the larger CD ratio. There must be some other kind of optic atrophy. Remember earlier that I mentioned that this patient had a history of blunt trauma to the right eye. We will often

Figure 3 [Top] Stereo photographs of another patient OD showing a pallid temporal optic nerve rim. [Bottom] Stereo photographs OS, same patient, showing healthy nerves with no pallor.

Figure 4 [Top] OD of another patient with temporal rim pallor. [Bottom] OS optic nerve head with healthy rim tissue in all quadrants.
see patients with pallid nerves, and quite dim vision, in patients with a history of traumatic optic neuropathy. In fact, you expect their vision to be quite bad. Could it be compressive optic neuropathy? Maybe. What about inflammatory optic neuropathy? It could be that, also.

So let’s do a couple of tests. What tests came to my mind at the time? I thought, “Let’s do a visual field. And how about an OCT?”

Let’s do a comparison here – 10 years ago, from when we last saw him in 1998, he had a reasonably good visual field for that 0.75 nerve. (Fig 5 [Top]) But then, man, look at that change on Fig 5 [Bottom] – do we expect that bad of a visual field from a 0.75 nerve? Typically not – that is generally reserved for more excavated nerves. And look at how diffuse that is. That’s not necessarily a glaucomatous finding until its more end-stage. Again, a 0.75 nerve is not yet end-stage.

Let’s move on to the left eye. (Fig 6) You will see why we were treating for glaucoma in 1998 – he had an inferior arcuate defect, getting close to the center of fixation. But then, interestingly enough, 10 years later, he’s got this sort of diffuse look, but if you look at the grey scale, it’s much, much worse temporally in the left eye. There’s a lot of zeroes, a lot of missed spots, and that defect is respecting the vertical midline pretty well.

We have a diffuse visual field loss OD, and temporal field loss OS. Any time you hear temporal visual field loss, especially vertically respecting the midline, you need to start thinking chiasm. This is concerning for a patient. Why are we not seeing the right eye with temporal field loss? Well, we are, but we are also seeing it with nasal field loss.
So let’s think about what could cause that. It does not look glaucomatous to me. It could be a compressive lesion. What about a stroke? That is highly unlikely, because we would have to have a left homonymous hemianopsia and then somehow explain the right visual field loss of the right eye. I think that’s a lot more challenging to do, so I don’t think it’s a stroke in this case. It could be a compressive lesion, which is, in fact, what it is.

Look at this patient’s MRI, shown in Figure 7. This is as if you are looking straight ahead at the patient, and they are looking at you. The left side of the MRI is the patient’s right side, and vice-versa. Right there, smack dab in the middle, is the patient’s pituitary. There is a huge lesion there, and not only is it there, it’s actually pushing forward a little bit and over to the side, more to the patient’s right.

Figure 8 is more of an ‘under the patient view,’ as if you were looking upwards towards the patient. In this patient’s example, this pituitary tumor is extending more towards his right optic nerve. This is what can happen with pituitary tumors. This patient’s tumor is not only expanding in more of a concentric fashion, it’s not expanding like a regular round ball; it’s actually moving forward and towards the right a little bit. This is what we get concerned about with these tumors – they can spread and cause a bitemporal hemianopsia, but if they push forward to one side or another, they can actually compress on the optic nerve. That is what happening in this patient – he is getting compressive optic neuropathy in addition to his pituitary tumor.

He had quite a large pituitary adenoma (2.5 x 1.6 cm) and this is called a macro adenoma when it’s this large. At the time that they diagnosed this, the decision was made to monitor this patient only. Honestly, looking back in hindsight, maybe it was the right decision. This patient is dead now. I don’t think it is from this tumor, but as you saw, he was pretty sick. He kept telling the neurologist and the neurosurgeons that he wasn’t really having any vision loss or changes, though his right eye was pretty much non-functional. I don’t know why the decision was made, but the patient passed away with some pretty good vision, and he was relatively happy with the way things were, so maybe it was the right call. Looking back, it’s hard to say, “Maybe we should have done a resection of that.”
**Pituitary Tumors**

There are different types of pituitary tumors. There are hormone-producing and hormone-inactive. Depending on the size, we may call them macro or micro adenomas. If it’s a micro adenoma, we are probably just going to normalize their function and reduce the symptoms and signs of the tumor. A lot of times in a micro adenoma, this is done with just medication alone. If it’s a larger adenoma, a macro adenoma, they are going to do a surgical resection of that. Surgery can be done transsphenoidally up through the nose, or transcranial where they actually cut open the cranium and go in. By and large, I’d say it’s largely done via transsphenoidally to avoid the more invasive procedures.

The take home on this patient is that you really need to check both eyes. There are two reasons for that in this patient; the first is that his good eye is the left eye, and we want to keep that good eye ‘good’. He had a 0.9 nerve, and is more at risk for losing vision in both eyes, which would be profound. The other reason is that it actually gives us clues as to what is going on. When you see dense visual field loss on one side, it’s very important to check the other eye, especially when one eye has that dense field loss, and the other eye is temporal, then you have that junctional scotoma as we’re seeing here, which is suggestive of pituitary complications.

When you have pituitary changes, they say that you will get different things. One is that, such as in this patient, you can get atrophy in the form of pallor as opposed to excavation. The nerve can only be damaged, or at least manifest that damage, in so many ways. One way is excavation as we see in glaucoma and some other optic neuropathies, but you can also get pallor, which is something you will see in patients with pituitary tumors or other forms of compressive optic neuropathy.

We also have to recognize what is pallid compared to what is excavated, and focusing on the remaining rim there will really help us. We’ve been fooled plenty in our clinic, and we see a ton of glaucoma. Sometimes we have to look a little bit more carefully and say, “That’s not glaucoma. That’s actually pallor.” We also have to know that glaucoma typically doesn’t cause pallor.

What is the urgency on a patient like this? If we are talking about brain tumors, we probably want to get this resected sooner rather than later, or normalized sooner rather than later. Typically we are referring this to neuro surgery and/or neurology, or endocrinology. It depends on what kind of practice you have – you may be referring this to a primary care provider if you need someone who can get them in the hands of the right people. Or, you may be referring to your local ophthalmologist if you are more comfortable with them taking care of this.

**Patient 2**

Patient number 2 is a 59 year old Caucasian male. He’s got an extensive medical history including hypertension, COPD, arthritis, kidney failure, anemia, hyperlipidemia, carotid artery stenosis, and he’s a bilateral amputee with peripheral vascular disease. That is actually why he is an amputee – it’s because of the peripheral vascular disease. Keep in mind, this is a 59 year old patient. He is already on Warfarin, HCTZ, Norvasc, Lopid, Metoprolol, Lisinopril, and Simvastatin. He’s a bilateral amputee who has a lot of things going on for one guy.

He comes in with a history of blur for 1 month. BCVA OD is 20/100, and OS 20/40. He’s got an APD. If you’re like me, you’re looking at this and saying, “This patient is 20/40 and 20/100, he just came in after
a month?” He’s got a lot of stuff going on, so I don’t know if he just didn’t think this was a big deal or if his vision was going to come back, but here we are.

Pressure was interestingly kind of low on day one, 6 and 10 mmHg. The anterior segment noted some mild cataracts, certainly not bad enough to contribute to this vision.

We look now at the posterior segment and we are seeing a sclerosed central retinal artery, with mild ONH pallor, but the retina appears perfused. Figure 9 is not him, but this is somewhat similar to what his eye looked like that day. The central retinal artery was pretty minimal. In regards to A/V ratios, we are looking at a very odd A/V ratio here. However, all-in-all, the retina looks pretty healthy. The nerve appeared a bit pallid but there was no whitening or cherry red spot that we would see with a CRAO.

There are six things that changed the way I practice after I saw this patient. I saw this patient pretty early on after I started practicing, so I just wanted to walk you through some of the lessons I learned so maybe you can avoid some of the pitfalls I fell into.

Number one is what is the “classic” presentation of a CRAO? We always talk about this whitening of the retina with a cherry red spot. This can be a bit misleading – it almost makes it sound like the problem is at the macula, when in fact the macula is looking like it normally does with that deep red appearance. It’s the rest of the retina that is sick, is not perfused. We get this swelling of the nerve fiber layer that causes a diffuse cotton wool spot, if you will. About a month or so after this, once the retina looks re-perfused, then it doesn’t look so sick anymore. It doesn’t look white or pale, it can look normal. We start off with this pallid, sick looking retina with a cherry red spot. (Fig 10) Figure 10 Right is a pretty cool photo, isn’t it? It has that segmental blood flow going through the arteries there.

Once the retina looks re-perfused, this is a difficult diagnosis sometimes. But you will see on a patient like our patient, it’s easy to see that there are fewer arteries coming out of the optic nerve, and it’s harder to see them. The A/V ratio is way off.

The differentials in this patient include a CRAO, an ophthalmic artery occlusion, ocular ischemic syndrome, and giant cell arteritis.
The reason we are worried about Giant Cell Arteritis, or Temporal Arteritis, is that 5% of CRAO’s are from this. This number, when I first learned this, was 10%. There’s a wide range of numbers that you’ll see on this in the literature, but the number was always 10%. It was from some study out of Bascom Palmer years ago, but honestly when I look back, I cannot find that reference for the life of me. Most of the other numbers tend to hover at around 5%. If we don’t know this, and we aren’t looking for this, there is the potential that the patient could have both really bad systemic complications, and/or really bad ophthalmic complications. Thus, you really have to pay attention to this any time you have a patient with a CRAO. I think you are obligated to do testing for GCA. At the very least, you need to ask the patient about symptoms suggestive of GCA.

In this patient, we did a little bit of extra testing. (Table 1) His ESR is slightly elevated, but keep in mind that he’s got a lot of other stuff going on. His C-reactive Protein was normal. The only thing that was a little bit off was his blood cell levels. He was anemic, and you should keep that in mind. That’s one of the reasons for checking the CBC when you’re looking at GCA. Any sort of elevation in the red blood cell or white blood cell count can throw off your ESR. Thus, it’s important to do the CBC with all of that.

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<td>ESR</td>
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<tr>
<td>CRP</td>
<td>0.5 (normal)</td>
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<tr>
<td>CBC</td>
<td>Abnormal RBC, HCT, HGB (Anemic)</td>
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<tr>
<td>Carotid Doppler</td>
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<td>Carotid Angiography</td>
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We also did a carotid doppler and carotid angiography on this patient. The carotid doppler results are in the text box on the left. As you can see, he has had a history of a stent in his right common carotid. It looks to be somewhat collapsed, though. That is the same side that had the artery occlusion.

A month later, which is 2 months after the start of his blur, he comes in with a very sluggish pupil and now his vision is hand motion. Thus, we have progressive changes.

Five months after all of this, he comes in with a dense, hyper-mature cataract. He went from having mild cataracts to an opaque white cataract that you just cannot see through. He presents with neovascularization of the iris and neovascularization of the angle for 360 degrees.

Lesson #3 is maybe this is further back than just the central retinal artery. When we see a CRAO, it’s not easy to know if the occlusion is further back. Could it be I the ophthalmic artery? Could it actually be all the way back into the carotid artery like this patient had? I’ve given this lecture before, and people ask me afterwards, do I think that this patient had Ocular Ischemic Syndrome (OIS)? Probably, but at the time I really didn’t know and it’s hard to tease that all out, to determine if it really was OIS, because this isn’t a typical OIS presentation. It wasn’t like he came in with mid-peripheral hemorrhages. He came in with what looked like an older CRAO. It was almost in retrospect that we were able to see the changes over time that we were able to realize that this was probably something bigger than just a CRAO. There is one of my lessons.
Another lesson is that CRAO’s can develop anterior segment neovascularization. The rate on this is unknown, but you’ll see literature state that it’s anywhere from 5 to 15%. I have seen three patients with a CRAO who have developed this, and that seems unusually high, so if you have a CRAO, don’t come to me because you’ll develop anterior segment neovascularization. Just kidding. But it’s one of those things that it’s not common, but interestingly enough, I’ve seen it a few times. This patient did develop it, and we ended up treating the neo without much success.

The interesting thing about this is that you would think that people who would develop neovascularization would be those with artery occlusions, more so than a vein occlusion. Artery occlusions are less likely to develop anterior segment neovascularization than a vein occlusion. It must be that a CRAO is a dead retina, where a CRVO is a sick retina. The sick retina sends out the VEGF much more than a dead retina? I’m not certain if that’s the answer or not, but it’s an interesting thought.

We treated this patient with Avastin for his neovascularization, which didn’t really do much for him. In addition, we decided that, since he wasn’t in any pain, to just watch that eye. We also decided to not treat the cataract, either, unless it became more of a problem.

Another lesson in this is that these patients with a history of artery occlusions are at greater risk for a stroke and heart attack. In fact, years ago when Hollenhorst and Offenbach did their original trials looking at asymptomatic retinal emboli, they found that patients were more likely to develop a heart attack than a stroke. That’s interesting, right? One would think that in a patient with an embolus in the retina, presumably from the carotid artery, would be more likely to have that embolus travel not only to the eye but also to the brain. But what we know is that they can also come from the heart is well. Thus, it is very important for a patient with a CRAO to get both a carotid artery study and an echocardiogram.

As we are looking at these patients, there have been rule changes as to when we are supposed to refer them. This is pretty new stuff, at least to me. If a patient has an artery occlusion, they are considered to be a high stroke risk and need to be seen today in the ER. Now they are considering the eye as an extension of the brain, which makes sense – the eye is a card-carrying member of the central nervous system and should be included. When the eye has an ischemic attack, that is similar to the brain itself having an ischemic attack, which puts the patient at a higher risk of having a stroke. These patients are put under a stroke protocol in the ER, and are often hospitalized, which seems pretty aggressive, but there is literature out there that is backing the practice up. This is all recommended by the American Stroke Association and the American Heart Association. They think these patients need to be seen sooner rather than later.

The other group is patients who have amarosis fugax. These patients need to be seen today, as well. Especially if you are certain that it is transient visual loss, and not something else like a visual aura.

The one that is a little bit complicated is the patient with an asymptomatic retinal embolus. Do they need to be seen today? Or is that something that can wait? The way I’ve been handling this is that I give some urgency to it. I let their PCP, their cardiologist, or their neurologist know, and give them the head’s up as to what’s going on, and they can make the call as to the urgency. A lot of these patients that I’m seeing with these have a long history of complications from it, and they are already on blood thinners like a high dose of Aspirin, warfarin, etc.
Last but not least, is compassion. The day I saw this patient was a very busy day at the clinic. I was working with a student doctor, and I really did not have much patience. I ended up seeing this patient whom I see quite often after-the-fact; he is a very memorable patient. I still see him in the hallways, and he is still alive many years later. That day, I was just off, and did not have much compassion for that patient. This is something that really stuck with me ever since. He had just lost an eye, and potentially had some very serious systemic complications and I just didn’t feel like I was very present for him that day. I kick myself for that every time I think about it. Hopefully you can avoid my pitfalls and be there, and have some compassion for our patients, even if they are there for minimal things. Especially for big things like this.

Patient 3

Let’s keep rolling and talk about patient number three. This is a 58 year old black male who comes in complaining of blur up close. He is myopic, with not too bad vision: 20/20 OD, 20/25 OS. His anterior segment was all good, no APD, and good pressure (IOP 17/17). Everything was really panning out OK for him. Then I look in the back of his eye. Honestly, if you and I are looking at Figure 11 (top) together, we can be honest and say there is not a lot going on here. The exposure of this photo makes it look like his nerve might be pallid – it is not. His macula looks a little atypical and that is what you want to focus on. Take a few seconds to pause and look at his macula and I think you will agree that it looks a little odd. Figure 11 (bottom) is what his OCT looks like. What is that OCT remind you of? There is a gap there in the photoreceptors that is missing.

Figure 12 (left) is the patient’s left eye, and honestly, the exposure did a really terrible job of showing the macula. Let’s look at his OCT. (Fig 12 right) Really, there is not a lot going on here, but there is some disruption in the exact same spot as earlier. This is a classic OCT presentation of solar maculopathy. His right eye almost looks like a stage 2 macular hole. Maybe there’s a horseshoe or semi-circle “C” look to it. You will see that
many times in these thermal burns. That’s exactly what the OCT looks like right there. Thus, our diagnosis is solar maculopathy.

What is interesting about this is that the OCT can really guide us in this. I think historically these patients may have gotten diagnosed as amblyopic, or maybe we just wrote it off as “not too bad of vision” or atypical macula. Now, we can exactly pinpoint what is going on with these patients. Not that there is any treatment for it, but we can at least pat them on the back and tell that that, prognostically, this is not going to get worse.

This is a thermal burn. A patient with a psych diagnosis is really key here. A lot of times, they are stargazing, if you will, but in this case, sun-gazing, as they have certain psych diagnoses where they might be doing that more. If there is a solar eclipse, not a lunar eclipse but a solar eclipse, then these patients can get this because they were staring at the sun. Essentially what is happening is that the cornea and the lens of the eye are hyper-focusing that heat onto the macula, and it’s simply burning it. Some patients who have a history of drug use, specifically psychotropic drugs that would make you see the sun as “crazy things,” such as LSD, or something like that, might be looking at the sun for an extended amount of time.

This patient had adjustment disorder, a history of different dependencies, alcohol abuse, so he fits that profile just a little bit. Honestly, I’m just straight-up asking these patients, “Hey, have you ever looked at the sun for an extended period of time?” They are usually pretty honest with me. Sometimes they even come in telling me that they have done that historically.

You will see in Figure 13 (Left) sort of a magnified view of what that looks like if you crank up the magnification in your slit lamp. There is a nice little burn there right in the center. These guys usually do pretty well. A lot of times they have trained themselves to eccentrically view things. Figure 13 (Right) shows a very high-definition OCT of that burn. It’s pretty cool to see that gap so defined. It’s as if someone just took a laser and took a chunk right out of the retina there.

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<td>Adjustment disorder</td>
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<td>Alcohol Abuse</td>
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<tr>
<td>Inadequate Housing</td>
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<tr>
<td>Depressive Disorder</td>
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<tr>
<td>Tobacco Dependence</td>
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*Figure 13 [Left]: Magnified view of a thermal burn in a patient’s macula. Figure 13 [Right]: High-definition OCT scan of the thermal burn in our patient.*
Patient 4

Patient 4 is very interesting, and we have seen her quite a bit in our clinic. She is a 30 year old white female who complains of headache and vision loss. She has had a history of vision loss in the right eye and double vision, but it is improving now. She is currently on Diamox and Butalbital, which can be for generalized headaches.

This one is kind of easy on our end. She actually came to us with a diagnosis. She has 20/40 OD, CF OS, an APD OS, and as you can see from the table, the left eye overall appears to be the worse eye.

![Stereo views of Patient’s nerve OD and patient’s nerve OS.](image)

What are our differentials? Papilledema could be a differential, and it could be from intracranial hypertension, it could be from malignant hypertension, or it could be from a space-occupying lesion. Could it be from optic nerve head drusen? How about uveitis? What about optic neuritis?

It wasn’t drusen. The patient did not have uveitis, nor did she have optic neuritis. This bilateral presentation is not typical for that, nor did she have pain on eye movement. In addition, CF vision is pretty bad for an optic neuritis. You can see it sometimes, but it’s not usually quite that bad. Thus, we are going with papilledema, which is bilateral nerve swelling from increased intracranial pressure.

These patients can get this from intracranial hypertension, which sometimes can be called pseudotumor. They can get malignant hypertension, which is very, very high blood pressure, we are talking 200/140, not your 150/90’s. Space-occupying lesions, something pressing in on the brain, can also cause it.

We took some photos of her eyes. Figure 14 (5 & 6) shows her right eye. If you have a stereo viewer you may be able to look at this, but this is a swollen nerve. You can see that whitish area, which is a demarcation line for where the edema has been.

Fig 14 (9 & 8) show her left eye. You will notice here that there is sort of a demarcation line in this eye that is moving out and is getting better. It is so hard to show optic nerve edema in 2D like we are right here. You really need 3D, which is why we took these pictures in stereo.

Look at the color of the nerve OD and the color of the nerve OS. There is quite a bit of difference; the temporal nerve OS has quite a bit of pallor.

<table>
<thead>
<tr>
<th>Table 7</th>
<th>Patient 4 Test results</th>
</tr>
</thead>
<tbody>
<tr>
<td>BCVA 20/40 OD, CF OS</td>
<td></td>
</tr>
<tr>
<td>APD OS</td>
<td></td>
</tr>
<tr>
<td>Confrontation VF Full OD, Limited OS</td>
<td></td>
</tr>
<tr>
<td>Color Vision: 10/10 OD, 1/10 OS</td>
<td></td>
</tr>
<tr>
<td>Anterior Segment: Unremarkable</td>
<td></td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Table 8</th>
<th>Differentials</th>
</tr>
</thead>
<tbody>
<tr>
<td>Papilledema: Intracranial HTN</td>
<td></td>
</tr>
<tr>
<td>Malignant HTN</td>
<td></td>
</tr>
<tr>
<td>Space Occupying Lesion</td>
<td></td>
</tr>
<tr>
<td>ONH Drusen</td>
<td></td>
</tr>
<tr>
<td>Uveitis</td>
<td></td>
</tr>
<tr>
<td>Optic Neuritis??</td>
<td></td>
</tr>
</tbody>
</table>

![Stereo views of Patient’s nerve OD and patient’s nerve OS.](image)
This patient is 62” tall, 462 lbs. She has a BMI of 85. I remind you that normal is hovering around 20, typically. Thus, she is quite overweight. Her blood pressure that day was normal (110/80). You have, have to, have to check the blood pressure on these patients. With bilateral ONH swelling, you could have malignant hypertension, which is a medical emergency, they need to be seen that day, that hour, as soon as you can to get their blood pressure down. They are at risk for some big time complications, so if you don’t measure the blood pressure you are not going to find it.

We were going to do an MRI on this patient. We did a lumbar puncture (LP) on her, and it was 490, then it went back down to 310. Keep in mind that opening pressure in these patients is usually around 250 mmH2O in obese patients. It is different in obese patients than it is for those who are not overweight.

A month later, her MRI was normal, her vision was a little bit better in her right eye (20/20 OD, 20/200 OS), and a little better OS. She had had a VP shunt, and you may be saying to yourself, “I’m not really following here... she came to you with this vision, she came to you on Diamox, what is up?” What happened was that she was at a different VA, and she had already been diagnosed with Intracranial Hypertension, and what she ended up doing was coming to our VA before she had the chance to receive treatment there. Now she’s showing up in our clinic saying she needed help. They ended up doing what is called a ventriculo-peritoneal shunt, shunting from her ventricles all the way to her peritoneum, her abdomen area. That helped the pressure go down, along with Diamox. Her right eye is mildly swollen, her left eye is swollen but it is atrophic.

Figure 15 [Top]: Visual Field OD, showing mild field loss. [Bottom]: Visual Field OS, showing profound loss.
Let’s look at her visual field. (Fig 15) There is some mild loss OD, but not too bad. Her left eye, however, was quite bad.

Now we have a lot of interesting information. 8 years later, she is not on Diamox, but her VP shunt is working well. She has minimal visual field loss OD, and her visual field loss OS is stable in 2012. It looks a lot better than it did back in the day, but we still have some visual field loss.

So why is the one eye 20/20 and the other eye 20/200? This all has to do with sick vs dead nerve fibers. When we see a patient with bilateral swollen nerves, we don’t know if those nerves are just sick, or they are dead. In this patient, we have a pretty good idea even from day 1, that her left nerve has some dead fibers, right? It had some atrophy, and it was pallid. But once we do the VP shunt, or once we put them on Diamox or they lose weight, we really don’t know if this patient is someone who will get their function back, or is it gone for good? Once we do the treatment, we will have a better idea. In her case, the nerve fibers returned to health in the right eye, but in the left they were dead. This is much, much different than we were taught, right? We were taught that these patients get bilateral enlarged blind spots, and that’s about it. But it’s a lot worse than that – if the edema is there long enough, it can compress on the optic nerve fibers, the nerve can atrophy, and then we get problems like this patient had.

Figure 16 [Top] Visual Field 8 years later, OD. [Bottom] Visual Field 8 years later, OS.
She has Idiopathic Intracranial Hypertension. We did an MRI, and nothing came of it. She had elevated cerebrospinal fluid pressure upon her LP, and we were able to evaluate and make sure she didn’t have any other serious complications. She has this, and it used to be called Benign Intracranial Hypertension or Pseudotumor Cerebri, and I think a lot of people still call it Pseudotumor. However, most people are moving towards this Idiopathic Intracranial Hypertension (IIH).

We know IIH happens frequent enough to see, but it is not terribly common. It is usually in young women, who are overweight. We know that if you are 10% overweight, you are 13 times more likely to develop this. The causes, however, are unknown. We are not really sure why high CSF pressure happens in people who are overweight. Is it something to do with different hormonal regulation? Why does it happen in women? We are not really sure.

The symptoms can be varied. A lot of times, these patients are asymptomatic but we stumble upon it. It’s not like bilateral enlarged blind spots are something that people notice very often. When they do have symptoms, it’s typically headache that can change with posture. It’s usually described as a generalized headache all around. They can have blurred vision sometimes, especially if there is enough edema that it is spilling over into the macula. Or, in the case of our patient, there’s enough edema that it’s actually damaging the optic nerve.

They can also have diplopia. More often than not, that’ll be the 6th cranial nerve – they will have a lateral rectus palsy. The reason why is cranial nerve 6 actually travels along our temporal ridge where the bone is, and if we get edema, it presses the nerve up against the skull and damages it that way.

Sometimes these patients can also be nauseous.

This is a diagnosis of exclusion. We want to do MRI’s, lumbar punctures, and as I mentioned earlier, you have to measure the blood pressure.

The treatment for this is varied. These patients will often go through weight loss. If you get as little as 10% weight loss reduction, you can often times reduce the course of this. In a patient who is 460 lbs, like this patient, that’s 46 lbs. That is a lot to lose. I don’t want to be so flippant about it, like “Oh, just lose 10% of your weight.” In someone like me, that is quite a lot. I don’t think I could lose 10% of my body weight very easily. Diamox can help, but it’s difficult because that medication comes with a lot of side effects. Surgically, we can also do a VP shunt.

Interestingly with the weight loss, what is more dangerous here? Is it more dangerous to do a bariatric surgery like a lap band or stapling the stomach where you may not be able to get as much food and you can lose weight that way? Or a VP shunt? What is more dangerous?

Patient 5

I have a 64 year old white male who complains of blurred vision since last year. He is positive for diabetes. His vision is 20/25 in the right eye, and 20/40 in the left. Everything in the front part of the eye is normal.

Figure 17: OCT from OS of Patient 5.
But then we look at the back of the eye and we see this OCT (Fig 17). What does this remind you of? You might think maybe VMT, maybe ERM, but it’s missing tissue there. That is not what those two conditions do. Could it be a macular hole? It doesn’t go all the way down to the RPE. Look at Figure 18. It looks like VMT, but it has these two large pockets. Are those cysts? Or what is going on here?

This is an impending lamellar macular hole, and I want to discuss what we do with these later. What I’m going to be doing for the next quite a bit of time is I’m going to walk us through some different macular diseases with OCT, what we are looking for, and how we go about fixing these.

**Patient 6**

I have a 74 year old Hispanic male with blurry vision. 20/40 OD, 20/25 OS. He’s got a cataract, so maybe it’s that? But then we look at the back of the eye, and we see this. This is a classic lamellar macular hole. Maybe we’ll call this a “reverse-anvil” look. It’s a hole of the macula, that does not go all the way down, full thickness, to the RPE. That’s what a full-thickness hole does – it goes all the way down to the RPE.

**Patient 7**

This is a 65 YO White male with progressive blurring. This guy is near and dear to me – we talk all the time and I love seeing him, but I hate what is going on with his eyes. He started out with me in 7/2014 with not too bad eyes. 20/20 OD, 20/25 OS, mild NS OU, and a mild ERM OU. Four months later, he is 20/20 OD and 20/50 OS. He’s got a mild ERM OD, and a moderate ERM OS. Let’s fast-forward to 2/2015. Now his right eye is starting to get worse. He’s now 20/30 OD, 20/50 OS with a moderate ERM/VMT OD and a moderate ERM OS. Fast-forwarding to August 2015 we are getting this huge progression: 20/50 OD, 20/200 OS with moderate ERM/VMT OD and moderate ERM OS. Looking at Figure 20 [Top], you can see how his macula looks. Honestly, you look at that, and you’re kind of surprised he sees as well as he does. This is his right macula, the 20/50 eye. Figure 20 [Bottom] is his left eye. There is quite a lot of
disruption there, right? I’m still wondering how he sees so well. That is an ERM just tugging and pulling on the tissues there. We’ll come back to him in a little while.

**Patient 8**

Now we have a 75 year old white male. He’s got blur when reading. BCVA OD is 20/40, OS is 20/30. PCIOL OU with the anterior segment being unremarkable. Look at Figure 21. We can see here the vitreous pulling right on the macula. This is vitreo-macular traction (VMT). This is a classic example of it. You know when kids draw seagulls on the beach in their pictures? This has a classic “seagull on a beach” look. That’s the best way I can describe it.

This guy has had a recent exam, and at that time was 20/20 OU. He was released without treatment at that time.

**Patient 9**

Patient 9 is a 75 year old white male. He has blurry vision OU, and has had cataract surgery OU. His health problems are limited, including COPD, DM and HTN. His anterior segment is unremarkable. BCVA OD is 20/50, and 20/100 OS. Looking at Figure 22 [Top], we see a full-thickness macular hole in his right eye. You’ll notice a few things about this: thing number one is it goes all the way down through the whole retina all the way to the RPE. It’s got some cystic changes in the remaining retina that is there, which is pretty typical of that fluid cuff around a macular hole. In this patient, you can see somewhat of an operculum above it – tissue attached to the vitreous above it. That’s kind of interesting. Figure 22 [Bottom] is his left eye.

Look at the top photo. This is the 20/50 eye – look how close together that is. Then look at the bottom photo – see how separated the edges of the holes are. Depending on how close these are together depends on the acuity. I was amazed that this patient saw 20/50, because I was taught most macular holes are 20/100, 20/200 or somewhere around there in that range, and I do think that’s true. In this patient, the vision is explained by the retina being close enough together.

**Discussion**

Let’s have a little discussion about ERM’s, VMT’s, lamellar macular holes and macular holes. Historically these patients were diagnosed by a combination of fundus appearance and visual acuity. What I mean is that sometimes an ERM can form what is called a “pseudo-hole.” It looks red, it’s round, and it looks a
lot like a true macular hole, but their vision is not nearly as bad. Usually ERM’s do not get as bad as macular holes. They tend to be much more in the 20/25 to 20/60, 70, 80 range. Whereas macular holes tend to be more in the 20/200 range. Lamellar macular holes are the same way. They look kind of like a macular hole, but it’s got a much better acuity.

Now we have OCT, and that’s really changed the accuracy of our diagnosis. This is one of the best examples of how to use an OCT. It gives us a much better idea of a prognosis, better guidance for treatment, better guidance for referral for when to treat, and the surgeons can look at this and give a much better idea of what is going to happen. We can also monitor them post-op.

An ERM is traditionally called Cellophane Maculopathy or Macular Pucker. What happens is that we get traction of the retina because when there is separation of the vitreous from the retina, you can get cells that migrate under the ILM and cause this sort of tangential traction. If you actually dissect the tissue that is in these, there are muscle cells there, which is why we get some of this traction. If there’s enough traction there, you can distort the shape of the vessels inside the retina and can get edema, usually cystic edema. Look again at Figure 20 [Bottom] – added again here for convenience. Look how much traction is there – it is just pulling on this tissue. It’s not just pulling on the X-Y axis, it’s also pulling on the Z axis, kind of tangential to the retina.

These patients will complain of blur, distortion, sometimes metamorphopsia. Again, they have a wide range of acuity, anywhere from 20/20 asymptomatic to 20/200 as in this patient.

What about progression? This patient progressed. Should we tell our patients who have ERM’s to watch out for the symptoms of this? Should we give them an Amsler Grid? I do not think so. In my practice, we see a ton of ERM’s, and I mean a ton. These patients are almost always asymptomatic. It’s very uncommon for us to see someone who progresses. Maybe you have a different experience that sort of colors your expectations of this. In my experience, this has been something that does not progress very often.

If the patient has reduced acuity, they are typically saying 20/40, 20/50 or worse, then they will treat these patients. This a lot of time depends on how symptomatic the patient it is, and how aggressive the retinal specialist is. They will often do a broad ILM peel, an ERM peel with vitrectomy, and often times these patients need cataract surgery if they haven’t already had it. The theory is behind this is that we are going into the back of the eye to do surgery, messing around with the back of the eye, mucking things up, it can make the eye develop a cataract. Anytime you have a patient who has had a vitrectomy or a membrane peel surgery, you should anticipate this patient will develop a cataract. Thus, not only is this patient in for one surgery, they are actually in for two surgeries. The surgeries are often very successful, but patients should have a tempered enthusiasm for the visual acuity to recover. Why that is, whether it’s just distorted retinal tissue that cannot be flattened out and resolved with surgery, I don’t know. But these patients often have less-than-perfect visual acuity afterwards, and usually the consent form of the retinal specialist is very specific about expectations the patient should have. Thus, you should have those same expectations as well, when you are giving your informed consent to the patient.
VMT

Vitreo-macular traction (VMT) is caused because the vitreous has very strong adhesions to certain parts of the eye, and the macula is one of them. Here is what I was taught: the nerve had the strongest adhesion, followed by the macula, followed by the vitreous base around the ora serrata. Well, if the macula has this really strong adhesion, as the vitreous is detaching, it doesn’t let go as easily and it can keep holding on. If the vitreous detachment is incomplete, we can get this vitreo-macular traction, and it pulls on the macula and distorts the shape of it. Thus, instead of having a nice flat macula with a foveal pit, we get this sort of taught-looking retina pulling up, as seen in Figure 21. In this patient, it’s so tight that the traction is pulling the macula up, leading to distortion of vision and distortion of the retina. Eventually in a patient like this, you may get resolution on its own, like the patient I talked about earlier, and the vision can return to normal.

Treatment for this almost always, in my experience, includes monitoring only. Because the vision is not terribly reduced. It can be significantly reduced, but in most of my patients with VMT, their acuities hover in the 20/40 to 20/60 range. You can go back and do a vitrectomy. There’s also a new medicine out there called Ocriplasmin (Jetrea), and this is an injection into the back of the eye to sort of induce a vitreous detachment, pull that adhesion away so it can even out the retinal tissue. They did a trial on this, and they found that 25% of patients improved with Jetrea. If they did the placebo, then 10% of patients improved. That’s a pretty good outcome, though only ¼ of patients improve, so we really need to weigh this against the complications, which can be many, in addition to an expensive injection. A lot of retinal specialists I’ve talked to are not enthusiastic about this medication.

Complications of VMT can include patients having conjunctival hemorrhages, photopsias, vitreous floaters, blurred vision and injection-related eye pain. And don’t forget the potential for endophthalmitis. The cataract rate for this was around 6% (9% placebo) and increased IOP was around 4% (5% placebo). Keep in mind, even patients in the placebo group had both of these, so from that standpoint, this is pretty safe.

Lamellar Macular Hole

Let’s move on to lamellar macular holes (LMH). This is not a full-thickness hole. It’s got atypical borders in that it doesn’t come down in a very clean, smooth way like you’ll see with a macular pseudo-hole. It also definitely doesn’t have that rounded-edge fluid cuff look that a true full-thickness macular hole does. A lot of times you’ll see this as an inverted anvil. The acuity is typically better in your patients with full-thickness macular holes, hovering around where ERM’s and VMT do. These tend to be very difficult to treat. Surgeons can do a vitrectomy or ERM peel, though they are not very successful very often.
Figure 23 shows an example of a classic LMH. It doesn’t go all the way down full-thickness, but it does have that “reverse anvil” look with atypical borders.

**Macular Hole**

Lastly, let’s talk about a macular hole. This is a full-thickness retinal break. As I mentioned earlier, acuity typically hovers around that 20/100 to 20/200 range, though this was proven wrong to me recently – it can be as good as 20/50. Though, again, I highlight typically 20/100 to 20/200. Typically, you have a pretty wide opening to that macular hole with a fluid cuff. In the patient in Figure 24, one of my patients I’ve had recently, who had pretty good acuity. I think he was 20/40 or 20/50. Look how skinny that macular hole is! I sent this to the retinal specialist jokingly asking, “What’s going on here? Is this something I’m missing?” He responded, “No, that’s just a really mild macular hole.” This is a retinal specialist with many years under his belt, and he said he hadn’t really seen many like this. Again, they are not typically like that.

Figure 25 [Top] is a very typical appearance of a macular hole. You’ll get that large, red, circular appearance with a fluid cuff. This is one example where funduscopically, this looks identical to the pictures in the textbooks. Sometimes textbooks will show you the textbook example of anterior segment neovascularization, where the whole eye is covered in neo. In this case, this looks like the classic picture of a macular hole when you look at it funduscopically. You will see if you look with stereopsis the fluid cuff surrounding it.

Figure 25 [Bottom] is what it looks like with an OCT. You see the cystic appearance of the tissue where the fluid cuff is. Again, it’s all the way down through the retina, full-thickness.

The way these patients are treated is they are given a vitrectomy. They will do a broad ILM peel, meaning the internal limiting membrane is peeled from arcade to arcade typically. If there is an ERM present, the surgeon will peel that, as well. They will then do a fluid-gas exchange, meaning they will remove the vitreous and put in a gas bubble.

Complications of this are, as I said earlier, with any sort of retinal surgery we think about cataracts afterwards. The other trouble is that the patient can get lack of closure – holes don’t always close 100% the first time. The other trouble is the patient has to stay in a face-down position. The face-down positioning on this is pretty interesting – there was a trial that just came out in 2014 that looked into
face-down positioning. They did a broad ILM peeling in one surgeon and 20 patients or so, along with an ERM peel if necessary, a vitrectomy and a fluid-gas exchange. Instead of having the patient lay face-down for a week, he had the patient look at a 45 degree angle down, as if looking at a kindle, smartphone or book. They did that for the majority of the day for 3-5 days. It was not the strict “lay down for so many hours of the day.” That surgeon had a 100% closure rate! That’s pretty interesting, isn’t it? This has yet to be replicated in other trials, but it is very promising. If we can prevent the patient from having to lay face-down for a week or so, or even just for a few days, that would be really nice. Especially if we can do some minimally invasive stuff in regards to their life.

We have reached the hour mark here, so what I want to do is stop here and thank you so much for your time and your attention. I want you to please feel free to email me if you have any questions. I will be happy to answer any of those questions. I hope this was interesting to you and I hope you learned a lot. I try to make it practical and interesting. Thank you for your time, and thank you for your attention.

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