Thank you very much for asking me to speak. I have to tell you I don’t think I have learned so much in a day for a long time, it’s been fascinating. When I was asked to talk about the use of OCT in children the – a kind of fear came over me because the problem is that I have often seen pictures of pathology and I’ve not really understood what I was seeing. And it’s only since I came to Pittsburgh that I started to get a real feel for the anatomy and the function of what the OCT was able to do. So I’m going to show you the sorts of things that we are seeing here and some of the things that I’ve seen in London and I’m going to really ask questions of the people who developed the OCT to help me understand better what’s going on in pediatric cases. I should say that I have no financial disclosure to make, I have absolutely no conflict of interest.

So the first thing is and I’m sure most of you know this, it’s more as a refresher is just to try and give a correlate between what you see on the OCT which is at the bottom and what you are seeing in the histological specimen as a correlate, and again the same thing, just to get an idea what layers we are looking at when we look at OCT, just a refresher for those of you in the audience.

So let’s talk about macular cysts. Alex Levine at Wills Eye published a paper recently which was very interesting, it talked about macular cysts occurring in the pediatric population and this was the first case that they discussed Goldman-Favre syndrome, and you can see that when you look at those fundus photographs you wouldn’t really guess that the pathology is of these enormous cysts occurring right at the fovea, and this explains why in this case in the paper the child had 20/60 vision in either eye and had been accused of malingering for 3 years. And it’s very interesting that some of
the cases I’m going to show you people said that the child was malingering and only when an OCT was done did they actually turn around to the parents and say look, we are very sorry, your child has a sight threatening condition.

How about this? This is the first, the first time I used OCT clinically and reliably was when we were looking at cases of X-link juvenile retinoschisis. Now this is from the paper that I described from Alex’s group but you can see here this is a huge schitic area of schisis, this is the peripheral schisis that you can see, the typical cartwheel maculopathy that they talk about and this what has now become a very familiar image but you know when these images first came out on the time domain OCT it was a revelation for all of us. And these are cases that I’ve dealt with again cases some of them, this one in particular had been the child had been told and the parents had been told the child is malingering, it’s a hysterical visual loss.

And while you look at this image and you go well it’s obviously something wrong here, there is something about examining children who are let’s say about 6 or 7 years old with an indirect ophthalmoscope and trying to get a good view of the macula. And you often don’t. You know the child is moving around, they’ve had enough by the time you are looking at their fundus and actually a photograph or an OCT examination gives you more information than you are getting from your actual clinical observation. And again this is the other eye of the same child and you’d say you know how come you didn’t manage to get that? Well I can tell you that this child was for want of a better word a spoiled brat and you, you couldn’t get him to do anything until you put him in front of
this machine and in fact one of the things that we developed, this is from (inaudible), we developed a little cartoon projection at a point where the child could have a look at it and we could get good fixation with the eye we were examining. And all of these are tricks and things that you have to develop if you are going to examine children well enough.

And again another case. So one of the commonest diseases that gets missed is Stargardt’s disease in the first decade of life. And this is a really nice paper if you want to look at it where they look at the classical sign on fundus autofluorescence that Tom Freyberg was talking about, and they tried to correlate the absence of the fundus autofluorescence with the abnormal fundus autofluorescence and the junction loss between the IS and OS. It’s a nice paper, it gives you a lot of information and that’s the classical picture that you might see on fundus autofluorescence.

So let’s talk about a case. A 7 year old boy with a 6 month history of blurry vision, right eye 6/36, left 6/18, N8 for near both eyes open, Ishihara 2/17, now the Flash VEP and the full field Flash ERG were normal, and you would expect them to be normal because the ERG is and the Flash VEP are summation responses for the whole retina. So you can have a macular problem and have a normal Flash VEP and a normal full field Flash ERG. What we didn’t do or what wasn’t done in this child was a Pattern ERG which would give you more information about the macula. So that again on the photograph I’m sure you’d agree that the macula looks abnormal and it has that beaten bronze appearance and again so Joel, I hate to ask you a question, but that shows us the kind of typical dark area that you’d expect to see on a fundus autofluorescence. But that’s an infrared picture. And if you
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KEN NISCHAL, MD

put infrared – hear infrared funduscopy and Stargardt’s there is nothing in the literature about it. And the only reason we did this was at the time when we were doing these as we are now at Children’s we wanted to acquire as much data at one sitting as we could. And nobody can explain to me, and I’m hoping someone in the audience might, why should an infrared funduscopy in Stargardt’s give you this dark area? That’s what the fundus autofluorescence looks like, but why are we seeing that on the infrared, because I can’t explain that. But it’s never been described.

And as I go on I will show you case after case where things that have not been described are being seen in children because all the basic work that was done in OCT quite understandably was done in adults. So if there is a keen eager resident and I know that we have at least one at Pittsburgh, I’m only joking, we have lots, this is a real field to make a name in because there is nothing in the literature about these cases.

So let’s talk about another case, a 10 year old, 20/50 best corrected vision acuity right eye, 20/30 best corrected visual acuity left eye. That’s the red free in right eye, left eye and it looks okay. I mean there is possibly an area right in the fovea here where you might say – actually yeah just there where you might say there is something like a – something funny going on in the fovea. Now here’s the next question, is that fundus autofluorescence normal or abnormal? You know when I first say this, and this case is fresh, it’s only 6 weeks old, well not the patient, the case, when I saw the patient I thought that they looked normal and then we looked at the cross-sectional – look at that. Is that normal?
We have – actually we just got a consult on a patient who has just that exact same image, an adult, looks – it could have been that person’s OCT and I mean it looks like the photoreceptors are absent in the foveola.

Absolutely.

You’ve lost the IS/OS junction, looks like things end at the ELM, and it would be interesting to know what AO, Adaptive Optics looks like in this patient.

Okay. So when I saw this it reminded me of a child I saw exactly 4 years ago in London. And I phoned up Dorothy Thompson who is the head of our Electrophysiology there, and I said, Dorothy, do you remember that child with the foveal bit that was missing? That was the word I used, bit. And she said yes. And I presented it at ISCEV, which is the International Society of Visual Electrophysiology, and there were 6 other people who had the same finding and we published the paper. Foveal cavitation as an optical coherence tomography finding in central cone dysfunction, only been published 3 months ago. And it’s striking that this is now an early sign of cone dystrophy. This child’s EOG which we did was normal, the Pattern VEP is abnormal, the Flash VEP is normal, and we are waiting on the Pattern ERG because we want to do a corneal electro-pattern ERG rather than skin.
But the point is this is – it’s a very nice paper and they have a range of patients from 7 years old up to 39, and they – and you can see how with the different cases the cavitation varies. And that’s extraordinary, that is the case that I remember seeing in London 4 years ago. But again you know here is things that we would never have imagined as being an early sign. I mean I wondered if that was normal or not when I first saw the case. Obviously that was abnormal but you wondered what on earth was going on. And if you look, in this group of patients that they looked at they had one patient that had a mutation in the Stargardt’s gene and one patient that had a mutation in one of the achromatopsy genes. So it’s not pathognomonic of any condition but it is a sign of early cone dysfunction. And I think that’s – I think that illustrates very nicely that we see stuff sometimes and I just don’t have a clue whether it’s normal or abnormal.

So what about the neurofiber disorder, there is a lot of work being done about the neurofiber layer in amblyopia, let’s have a look at this case, a 5 year old male, 20/200 in the right eye, 20/50 in the left eye, noncompliant patching and that’s his refractive error, plus 250 right, plus 1, minus 1 at 90 degrees in the left. And the interesting thing for me is that if you look at a neurofiber layer that looks possibly thin it’s the left eye and not the right eye. So all the data that we have or the couple of papers that have been published I just wonder how robust that data can be when you consider that the normative data for this machine includes the youngest person as being 18. So again we have no normative data for any of these machines in the really important age group and that’s the first 8 years of life. And if you split that into under 2s and 2 to 8 I think that the information that we will get is different. And again if you look at the optic nerve profiles it’s actually the left eye that looks smaller
than the right eye. So again it’s comparing it to normative data that is the wrong age and perhaps the wrong ethnicity, in fact I know it’s the wrong ethnicity.

So there is a lot made and I’m not going to go onto this long because I know that Dr. Bonhomme is going to talk about this, but here is a child with clear evidence of raised intracranial pressure, papilledema, the child we know has a hydro – I was going to say hydrocephalus but I have to say hydrocephalus otherwise people just hiss at me. And so here there was – here is what the optic nerve head looks like and then there is a recent paper that compares optic disc drusen on your left to what papilledema looks like on your right. Now there was a paper that talked very convincingly about the boot sign in optic nerve head drusen, and I thought it was the face of Jesus sign and some people thought it was the sign of you know Dalai Lama. What I’m trying to say is you can see what you want to see when you are trying to make a point that is not valid. And the issue – oh I’m glad you got that because you know American humor doesn’t always stick around with me.

So the point is that apart from this fact that you get these sort of lumpy discs there is not much you can really say, and there is another paper that’s been published and again you know I must admit I’d be interested to hear what Dr. Bonhomme says, I rely much more on the clinical features and ultrasound measurements of the optic nerve sheath diameter rather than OCT for differentiating optic – this drusen from swelling. And you know these are – I can show you picture after picture that have been published but when you’ve got a patient in front of you I would say I still rely on the old
clinical sign of increased disc swelling, vessels crossing the disc on the eye with the optic disc drusen and looking at the pulsation, venous pulsation of the nerve.

So I’ve always wondered you know for glaucoma progression which OCT do we use, and I’m so glad I’ve wondered that because from today, today’s lectures it becomes apparent that you know you probably need to stick to one machine, that going from one machine to another doesn’t help. But you have to be very, very strict about the parameters that you are looking at to make sure that you’ve got a good scan and again you come back to this issue that really I think we can look at progression in children more reliably than we can look at absolute measurements again because we don’t have any normative data. So I think it’s important that if we are going to use this new technology that we use it in a way that’s safe for the – for what we infer from it. And I think progression is probably the way that I would look at it, but there are lots of questions. For example, if I operate on a baby who has a cup to disc ration of .8 and I reduce the pressure, let’s say the child is 9 months old, I reduce the pressure significantly, I know that the optic disc ratio will reverse to maybe .2 or .3. What we don’t know, and what we want to try and find out is does the nerve fiber layer change? Are we seeing a genuine change in nerve fiber layer or just the metrics of what the scleral canal and the tissue within it are doing, which makes more sense? But again it’s stuff that nobody knows and certainly has not been published.

I want to show you 2 or 3 cases that I think again illustrate some very interesting things in terms of parent education and parent understanding. These are not great pictures but this is a child who came
in with what I was told was an optic neuritis in the left eye and it was recovering, so that’s the picture of the left eye. That’s the picture of the right eye. Now the picture is not great but you can tell both nerves look similar and when I said to the parents look the other eye also looks like it’s had some sort of event the parents were very non-believing. And then we took the pictures. This is his MRI scan when he presented with the optic neuritis to the emergency department and on that side he has a perineuritis. Perineuritis is very rare in an optic neuritis, but it’s very not common but it can be seen in Leber’s hereditary optic neuropathy. And when you did – when we did the scan even though we didn’t have appropriate normative data I could show the parents that the nerve fiber layer was thinned in both eyes, that whatever was going on had happened in both eyes and yet he’d only had a painful episode in one eye. A lot of people think that Leber’s hereditary optic neuropathy does not present painfully, and in fact the largest series 134 families published from Queen’s Square in London showed that 22% of acute presentations were with painful eye movements. So again here I’m using this as patient education for the parents, not for the patient, but for the parents.

So what about the cornea? Again very useful to help explain what’s going on, here is a child with Noonan’s syndrome and he was referred to me because he’d had ptosis surgery and the local ophthalmologist, this is here in – just outside Pittsburgh, said I’m worried because I think I’ve given him exposure keratopathy because he came back to me after a year and he has these scars. And you look at that scar, if you didn’t get a good look you might think it is exposure keratopathy but there are features about this that don’t look like exposure keratopathy. When you do the Visante OCT look at this lesion that is absolutely subepithelial and anterior stromal. And it turns out that there is –
there are 3 reports of anterior stromal dystrophy in Noonan’s syndrome. But this is the first time anybody’s imaged this and I wonder if there are other children with Noonan’s whose scarring is being mistaken for exposure and actually is a form of dystrophy.

So here is my last case, here is a child who had a traumatic cataract, big iridodialysis and you could see that there had been hemorrhage at the macula, specifically at the fovea. And I said to the mother before we took the child for cataract surgery I could just about get a view, I said look, I don’t think the child is going to see very well, we do need to repair the iridodialysis, get rid of the cataract but the child is not going to see very well. We did the surgery, we did the OCT and you know for a parent to be able to see what the problem is, the problem that this is normal and there has been a big hemorrhage and a rupture at the fovea, they understand completely that there is very little that you can do and why it’s happened.

So in conclusion for me the OCT technology is raising more questions than necessarily giving me answers but that’s a good thing. For me it helps me explain the situation to parents in a much more positive, not positive more comprehensive way than I could have done previously. So I think there is a lot – this is a field that really excites me because there is so much that we don’t know but there is so much that we can learn. Thank you very much.