

# News in Review

COMMENTARY AND PERSPECTIVES

## Novel Sildenafil Use: Lymphangioma

Following on the heels of a serendipitous discovery at Stanford University,<sup>1</sup> doctors at the University of California, Davis, Medical Center have successfully treated two cases of pediatric orbital

lymphangioma with oral sildenafil,<sup>2</sup> the medication best known for its treatment of erectile dysfunction. Stanford doctors had previously treated a young child with sildenafil for pulmonary hypertension, and they also found that the child's large intrathoracic lymphatic malformation had greatly decreased in size.

**New options needed.** Orbital lymphangiomas are rare congenital malformations of lymphovascular tissue that are unencapsulated, bleed spontaneously, and increase in size with infections. Their treatment can pose an intractable chal-

lenge to patients and ophthalmologists alike. "Many different interventions, including systemic steroids, drainage with sclerosis, and surgical debulking, can be used for patients requiring treatment," said Nandini G. Gandhi, MD, assistant professor of clinical ophthalmology at UC Davis. "But none of these has demonstrated consistent success, and nearly all require general anesthesia, which adds another layer of risk for children."

Treatment often requires an individualized approach, she said, with choices depending upon the location



**BEFORE AND AFTER.** (1) Patient at 5 months old; he received seven months of sildenafil treatment starting at 12 months of age. (2) Same child at age 2.

of the lesion, age of the child, and extent of its effect on surrounding tissues and structures.

**Two success stories.** The UC Davis team's first case using sildenafil was a 12-month-old boy with a lymphangioma involving the right orbit. He had not responded to aggressive surgical interventions, which included repeated sclerosing therapy and drainage of the lesion, said Dr. Gandhi. "The child continued to have recurrences of bleeding to the point where he couldn't even open his eye. We were concerned about the effect of compression

on orbital structures and vision loss secondary to deprivation amblyopia." After treatment for seven months with sildenafil, the child's ptosis and cheek swelling significantly improved. The patient has remained in remission since discontinuation of the medication—nearly a year.

The second case was a 12-year-old boy with right-sided orbital and intracranial lymphangioma. Although he had undergone multiple sclerosing procedures throughout infancy and childhood, swelling and elevated intraocular pressure had recurred. Given

the extreme swelling and lack of light perception, enucleation was being considered, but treatment with sildenafil was tried instead. Eight weeks of treatment significantly reduced his facial swelling, pain, and chemosis, allowed fitting with a cosmetic scleral prosthesis, and prevented the need for enucleation. The authors reported that three months after the patient began treatment, there had been no

recurrences.<sup>2</sup>

Although the drug's mechanism of action remains speculative, said Dr. Gandhi, it is thought that by relaxing smooth muscles, sildenafil may allow the collapse of the cystic channels that would otherwise become dilated and fill with blood or lymphatic tissue.

#### First-line treatment?

Sildenafil is well tolerated and represents an exciting possible innovation for the

treatment of these lesions, said Dr. Gandhi. "For patients who do not require urgent surgical intervention, it seems reasonable to attempt treatment with sildenafil before moving to more aggressive treatments, but these decisions should be made in close conjunction with the patient's primary care team."

Since many questions remain about sildenafil's role for this condition, Dr.

Gandhi looks forward to more studies, including results from an investigational pilot study underway at Stanford.

—Annie Stuart

1 Swetman GL et al. *N Engl J Med*. 2012;366(4):384-386.

2 Gandhi NG et al. *JAMA Ophthalmology*. 2013;13(9):1228-1230.

*Dr. Gandhi reports no related financial interests.*

## Smartphone Update

# Mobile Funduscopy

Just three years ago, smartphone images of the fundus were possible, but hardly practical. The clinician had to grasp a 20-D indirect lens in one hand and a penlight plus the phone in the other—and then hold everything steady while somehow tapping the phone's screen to capture an image.<sup>1</sup> Not surprisingly, the quality often wasn't good.

But a group of smartphone-savvy retina subspecialists at Massachusetts Eye and Ear Infirmary (MEEI) reported that further attempts to develop a funduscopy device have paid off.<sup>2</sup> Their modifications yield high-quality still photos.

**A solution for use worldwide.** The mobile-phone approach to funduscopy is expected to be particularly useful in settings where expensive imaging setups are unavailable or cannot be easily used, said Shizuo Mukai, MD, associate professor of ophthalmology at Harvard Medical School and the

senior author on the paper. Consequently, the group published its paper in an international online, open-access journal in hopes of reaching ophthalmologists who practice in remote and underserved areas of the world. Traditional funduscopy devices, said Dr. Mukai, "are expensive, and there are often difficulties with power supplies and repair/maintenance service."

Even where resources are more abundant, the smartphone method has the benefit of convenience, Dr. Mukai said. For example, with a Koeppel lens added to keep the corneal surface moist and the lids open, fundus photos can be taken in children under general anesthesia. Or an ophthalmologist in an emergency department could capture fundus photos and e-mail them to a more experienced clinician for consultation. "My fellow, Luis J. Haddock, MD, who is the first author on the paper, uses this in the



**iFUNDUS.** A few apps can turn a smartphone into a viable fundus camera.

ER all the time," Dr. Mukai said.

**What you need.** Dr. Mukai said the group overcame the limitations of earlier smartphone-based systems with the help of hardware and software upgrades to the Apple iPhone, which his team uses. These innovations include the following:

- **Light source.** Recent generations of the iPhone (4 and 5) have an integrated light source next to the camera lens. The feature also is available on other smartphones.
- **Video app.** A low-cost app (FiLMiC Pro, \$4.99, Cinegenix: <https://itunes.apple.com/us/app/filmic-pro/id436577167?mt=8>)—origi-

nally for digital moviemaking—allows the physician to independently control focus, exposure, and illumination during video recording. The latter is important to getting the best exposure, and it gives additional assurance of safety for the eye, Dr. Mukai said. Cinegenix has said it hopes to release a version of FiLMiC Pro for Android smartphones in early 2014.

- **Video still app.** Two apps can capture high-quality still images from an HD video (MovieToImage, 99 cents, DreamOnline: <https://itunes.apple.com/tc/app/movietoimage/id383313096?mt=8>; and Video 2 Photo, \$1.99, Francis Bonin/PacoLabs: <http://pacolabs.com>). Video capture also is possible with the iPhone's native software, with a screen-grab during video playback, but the quality suffers, the researchers reported. —Linda Roach

1 Lord RK et al. *Ophthalmology*. 2010;117(6):1274-1274.

2 Haddock LJ et al. *J Ophthalmol*. 2013; Article ID 518479. doi:10.1155/2013/518479.

*Dr. Mukai reports no related financial interests.*

## Retina Report

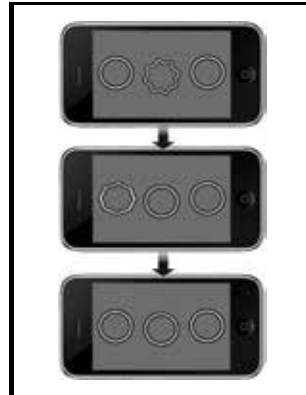
## Mobile AMD Monitoring May Ease, Improve Care

Timing is a key challenge in treating patients with neovascular age-related macular degeneration (AMD). Ideally, anti-vascular endothelial growth factor (VEGF) injections would be given right away, when leaking from abnormal blood vessels first occurs. But patients may miss the warning signs—distortion or blurring of vision—that often precede leakage. So physicians must rely on monthly checkups, which can be burdensome for patients and result in less-than-timely treatment. In ophthalmology and across medicine, physicians

are looking to patient self-monitoring as one way to boost efficiency and personalization of care.

Peter K. Kaiser, MD, a vitreoretinal specialist at the Cleveland Clinic, and colleagues followed 160 AMD patients aged 49 to 92 to check their satisfaction and compliance with a smartphone-based self-monitoring system—one component of a larger patient data system called the Health Management Tool (HMT).<sup>1</sup>

Patients were provided with smartphones equipped with two patient-input apps: myVisionTrack (mVT) and



**mVT APP.** The patient must choose the distorted circle before moving to the next screen, in which the circle is less distorted than the previous. The app then generates a value using the least-distorted circle the patient was able to identify.

Signs and Symptoms Questionnaire (SSQ), as well as a third HMT utility app that uploads mVT and SSQ data to a central HMT database, where clinicians can check patient-generated data.

Study participants used mVT to test their visual function over 16 weeks. Daily self-tests were completed by 80 percent of patients, and nearly 93 percent said the system was easy to use.

“Our results show that older patients can and will use a mobile handheld device to test their vision,” said Dr. Kaiser. “The HMT system accurately and consistently tracked patients’ visual function, even in those with poor vision. Physicians could easily monitor their patients via mVT data, and the SSQ offered timely feedback on any adverse postinjection reactions.”

—Mary Wade

1 Kaiser PK et al. *Retina*. 2013; 33(9):1863-1870.

Dr. Kaiser consults for Bayer, Chengdu Kanghong, Genentech, Novartis, and Regeneron, from which he receives grant support.

## Refractive Cataract Update

## Get Head Position Right in Keratometry

The expanding use of toric intraocular lenses (IOLs) has made meticulous preoperative astigmatism measurements even more critical to achieving good outcomes. Head tilt turns out to play a larger role in IOL outcomes than originally believed.<sup>1</sup>

“A small amount of head tilt can induce ocular torsional movement and maintain the astigmatic axis constantly,” said Choul

Yong Park, MD, PhD, associate professor in the graduate school of medicine at Dongguk University in Seoul, South Korea. “Because of this, most surgeons have not paid sufficient attention to the importance of correct head position when obtaining preoperative astigmatism data. But our study findings indicate that torsional compensation cannot always be expected and that strict control of an

upright head posture is very important in obtaining constant astigmatism data.”

To evaluate the effects of head tilt on keratometric measurement, Dr. Park and his colleagues examined the right eyes of 27 volunteers with a manual keratometer, an automated keratometer, and an IOLMaster (Carl Zeiss Meditec). Measurements with the manual and automated keratometers were taken in the upright head position. IOLMaster measurements were performed in five head positions: upright, 5 or 15 degrees of clockwise head tilt, and 5 or 15 degrees of counterclockwise head tilt. Some patients had an almost 10-degree change in the axis

of astigmatism with head tilt—a degree of axis error that can halve the beneficial effect of the toric IOL.

“I strongly recommend marking the midline of the patient’s face and making sure that the marking is exactly perpendicular before pressing the button of any biometric instrument,” Dr. Park said. “It is also important to check the astigmatism axis and to mark the surgical axis on the same day and at the same spot to minimize axis error.”

—Marianne Doran

1 Park CY et al. *Eye*. 2013 Sept. 20. [Epub ahead of print].

Dr. Park reports no related financial interests.



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## Bench Testing

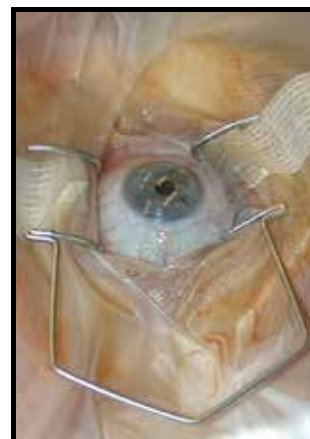
# Stiff Speculum Linked to Postop Ptosis?

Several studies have implicated the lid speculum as a factor in common postoperative lid malpositions after routine intraocular surgery. Recently, a group based in the United Kingdom examined forces exerted by lid specula.<sup>1</sup>

Cataract and retina surgeon Niall Crosby, MD, of Birmingham and Midland Eye Centre, City Hospital, Birmingham, said, “There is indeed a significant difference between two very commonly used types of specula,” specifically, disposable versus reusable models, which are stiffer.

“From our own observations, the most vulnerable patients are usually elderly, with a short horizontal palpebral aperture, where the speculum causes the lids to stretch into a tight square configuration. This is especially the case where a stiffer speculum is used,” he said.

The researchers undertook the study after oculoplastic surgeon Aidan Murray, MD, observed frequent postoperative ptosis in elderly patients with small horizontal and vertical palpebral apertures, and suggested a relationship with the use of a new, stiffer speculum popular with cataract and retina surgeons. Dr. Duncan Shepherd of the School of Mechanical Engineering, University of Birmingham, devised a way



**IN THE LAB.** The impetus for this bench study came from observations that the lid speculum may play a role in postoperative lid malpositions that can occur after routine intraocular surgery.

to test the specula.

Four disposable stainless steel and four reusable titanium specula were tested through compression. Stiffness in the reusable speculum was significantly greater ( $p = 0.002$ ).

“What surprised us was the magnitude of the difference,” Dr. Crosby said. He advises avoiding stiffer specula in vulnerable patients.

The team plans a clinical study to test variation in forces exerted on eyelids to determine correlation with subsequent lid malpositions.

—Laura Kaufman

1 Crosby NJ et al. *Eye*. 2013; 27(9):1098-1101.

*Dr. Crosby reports no related financial interests.*