

**SECONDARY GLAUCOMA:
Pseudoexfoliation (PXF), Pigmentary Dispersion Syndrome (PDS),
Neovascular (NV), Uveitic**

Many roads can lead to glaucoma. With the exception of primary open-angle glaucoma (POAG) and closed-angle glaucoma, there is a large group of glaucomas resulting from either genetic or physical conditions called secondary glaucoma. To discuss some of the more prevalent secondary glaucomas, on May 18, 2013, the Group welcomed Tania Tai, MD, Attending Physician, New York Eye and Ear Infirmary to discuss some of the more common secondary glaucomas – pseudoexfoliation (PXF), pigmentary dispersion (PDS), neovascular (NV), and uveitic glaucoma.

Unlike Primary Open Angle Glaucoma (POAG) Secondary glaucomas are characterized by an identifiable cause for high eye pressures.

PSEUDOEXFOLIATION GLAUCOMA

Pseudoexfoliation (PXF) glaucoma is one of the most common forms of the secondary glaucomas. A higher frequency of cases exists in colder climate countries such as Finland, Sweden, Norway, Ireland and Iceland. Russian Jews also show a higher prevalence. But warm countries are not exempt for it is common in the Middle East, Japan, and India.

PXF is also the most common cause of unilateral glaucoma although your doctor will watch your other eye closely. Since secondary glaucomas are associated with a physical condition, the cause of PXF has recently been linked to one defective gene (lysyl oxidase-like 1 polymorphisms), but medication to block the activity of this gene has not yet been developed. Not everybody who has pseudoexfoliation gets glaucoma; however, it is a systemic process affecting the skin and the visceral organs. While people with PXF live as long as the general population, they may be more subject to hypertension, stroke, myocardial infarction, aortic aneurysm and other heart diseases. In the eye, it is characterized by production and progressive accumulation of a fibrillar extracellular material that then occupies many ocular tissues especially the drainage channels of the eye. It may lead to both open-angle glaucoma and angle-closure

glaucoma and may also be associated with cataract, lens dislocation, impairment of the blood aqueous barrier, and poor dilation. Special care is needed with cataract extraction for the fibrillar material can load up on the zonule fibers that hold the lens in place weakening them and causing an instability in that region. If this occurs the replacement lens will need to be set in a section of the eye other than the lens capsule. PXF glaucoma can present with much worse high pressures, making progression more likely. As well, it tends to be more aggressive than primary open-angle glaucoma.

Management: In general the majority of the available medications depending upon the patient's sensitivity will act to lower the IOP, but with PXF glaucoma, more drops may be necessary. By and large your doctor will fashion a medical routine that will work best to maintain an IOP where vision loss is minimized.

Laser therapy (laser trabeculoplasty) is effective at first (It is speculated that accumulated material absorbs the laser energy better) but its effectiveness may drop within several years because the fibrillar fibers continue to clog up the meshwork. With laser trabeculoplasty, however, your doctor takes special care because of the higher risk of post-operative pressure spikes immediately following the treatment; nevertheless it can be an effective treatment and may be recommended by your doctor.

PIGMENTARY DISPERSION SYNDROME (PDS) GLAUCOMA

In Pigmentary Dispersion Syndrome (PDS) Glaucoma, the iris is bowed backwards with a concave configuration, causing it to rub against the zonules that release pigment. The pigment becomes dispersed throughout the anterior segment of the eye (the front part). It is deposited on the cornea, the lens and like PXF, the pigment material gets trapped in the drain causing malfunction in the outflow of fluid. Findings the doctor may see on slit lamp examination include a Krukenberg spindle (pigment deposit on the cornea) and iris transillumination defects. The transillumination occurs because light shines through the iris where pigment has been rubbed off. As with PXF not everybody who has PDS gets glaucoma. Usually, people with PDS are myopic and young. Although the syndrome is found in

both males and females, PDS glaucoma occurs more frequently in males.

As the trabecular meshwork clogs up, eye pressure rises and damages the optic nerve. People with PDS may experience sudden blurred vision, pain, and halos in vision due to rapid increases in pressure.

As the patient ages the lens in the eye naturally thickens and pushes the iris forward, reducing the rubbing against the zonules. In turn this reduces the pigment dispersion, decreasing the eye pressure. When this occurs, it is possible that the need for glaucoma medication can be reduced. As some patients age, they may find that they no longer need to lower their pressures, but the years of pigmentary accumulation may have a lasting effect.



Illustration of bowing backward.

Management: Medical therapy for PDS glaucoma consists of eyedrops to lower intraocular pressure, similar to those used in other forms of glaucoma. Miotics, such as Pilocarpine, may be particularly effective for it keeps the pupil constricted, reducing the amount of pigment released.

Laser: A laser peripheral iridotomy can be used to create a tiny opening in the peripheral iris that then allows the aqueous fluid to flow from behind the iris directly into the anterior chamber. This can reduce the concavity of the iris in PDS, and possibly decrease pigment dispersion. However, there is conflicting evidence regarding the efficacy of the procedure in reducing IOP in eyes with PDS.

Laser trabeculoplasty. Laser trabeculoplasty has been demonstrated to lower IOP effectively in eyes with pigmentary glaucoma. Caution must be exercised when setting the laser energy in this procedure, as the heavily pigmented trabecular meshwork in eyes with pigmentary glaucoma absorbs greater energy.

NEOVASCULAR GLAUCOMA (NVG)

Neovascular glaucoma (NVG) most commonly affects people with retinal ischemia, which can be caused by conditions such as diabetes, central retinal vein occlusion (CRVO), and carotid artery obstructive disease. It results from the formation of new (neo) blood vessels that are fragile, weak and abnormal. These blood vessels creep over the eye structures, including the iris and trabecular meshwork, growing over the drain causing it to malfunction and scar. Intraocular pressure increases and glaucoma can occur.

Management: Detection and treatment of ischemia early can minimize neovascularization and prevent the blood vessels from creating damage. Although intraocular pressures may not rise immediately, the doctor will want to treat the neovascularization. If treatment is begun early enough, the errant blood vessels growing over the drain can regress. Treatments to inhibit neovascularization include medications such as Avastin and Lucentis, and retinal laser therapy. If the neovascularization does not regress, scarring over the drain can occur.

Should pressures still remain high the standard medication drops are also used. Laser trabeculoplasty does not work for neovascular glaucoma.

UVEITIC GLAUCOMA:

Halos and light sensitivity are some of the problems uveitic patients face. Uveitis is inflammation of the uvea (the pigmented middle layer of the eye consisting of the iris and ciliary body together with the choroid). Inflammation in the eye can be caused by many different things, such as HLA-B27, syphilis, tuberculosis, sarcoidosis, rheumatoid arthritis, keratouveitis, herpes zoster and others. The list is long but your doctor will run a blood test to determine if you have markers for any of these conditions. It is possible to have uveitis, even without the conditions listed above. Uveitis can lead to high eye pressure because the drain can be affected by inflammatory cells, proteins, debris or fibrin liberated from a disrupted blood-aqueous barrier. The inflammation can also cause scarring over the drain, similar to that caused by neovascularization, which too can elevate

eye pressure. Secondary angle-closure glaucoma can also result in uveitic eyes. Angle closure with pupillary block occurs when inflammation in the anterior chamber causes 360 degrees of posterior scarring that blocks the flow of aqueous from the posterior chamber to the anterior chamber, resulting in acute angle-closure glaucoma.

Management: Steroids are used to treat uveitis, but long-term usage can also induce a rise in pressure. Doctors opt to use a minimum amount of steroid to keep the inflammation in check. Steroids have been reported to cause biochemical and morphological changes in the trabecular meshwork, decreasing aqueous outflow facility. At times medications other than steroids are used, but basically steroids are the first line of treatment. If control of inflammation with steroids does not bring down the pressure, the various pressure-lowering drops are used, similar to other forms of glaucoma. Laser therapy is contraindicated because of the ensuing inflammation following treatment and the prospect of additional scarring. The full panoply of eye drops are used, but if they do not control the pressure adequately, surgery is the next step. With uveitic glaucoma, however, surgery may not have as high a success rate as in other forms of glaucoma because all glaucoma surgeries rely on creating a pathway for the fluid to exit the eye, and that pathway needs to remain patent. In uveitis, because the eye is prone to inflammation, the opening created has a higher risk of closing.

We want to thank Dr. Tai for her most enlightening review of the above secondary glaucomas and to especially thank her for presenting this address in the Lunch Room under difficult circumstances. The Group is so grateful to learn more about the “secondaries” for many of us have one of these conditions.