Q:

- Patient comes to see you with this, and has IOP of 32.
- What is the Dx? The Ddx?
- Etiology?
- Epidemiology?
- How would you manage? Keeps walking you down the path to more aggressive therapy.
- Patient develops a cataract, how would you manage it?
Pseudoexfoliation Syndrome
—epidemiology
—age—70’s (rarely < 40 yo)
—increased incidence with age
—Framingham study 0.6% 52-64 yr, 5% 75-85
—25-50% of affected eyes develop glaucoma
—50% unilateral (but 25% become bilateral by 5 yr)
—sex

w/o glaucoma F > M

with glaucoma F = M

—rare in blacks
—common © Scandanavians, Navaho Indians
—steroid responder same incidence as general population
—DDx true exfoliation d/t separation of superf. layers of lens capsule after exposure to intense heat (eg, glass blowers), radiation -->
glaucoma uncommon
—ocular findings
—dandruff on lens capsule, iris, pupil border
—target pattern on capsule with central and peripheral zones of deposition separated by clear zone (from iris contact)
—pigment on ant. lens capsule
—depigmentation of pupil ruff ± transillum defects near pupillary border
—pupil dilates poorly (© iris hypoperfusion, microNV) b/c endothelium of bv’s also affected
—gonio—hyperpigmented TM
—narrow angles
—Sampolesi’s line (pigmented Schwalbe’s)
—weak zonules lead to
1—lens loss during cataract Sx
2—angle closure b/c lens can move forward
3—phacodenesis/iridodensesis
—“amyloidosis of eye” b/c
—weak staining with congo red
—intense staining with thioflavin T or S
—histology—exfoliative material from all epithelial derived structures in eye found on lens capsule, CB, iris pigment epithelium, subconj, skin, heart, liver, lung, kidneys
— course
— present with more cupping & field defects than POAG
— IOP fluctuate more --> need DTC, closer f/u (eg. q4 months)
— periodically re-gonio (at least yearly)
— will respond well to ALT (best of any glaucoma)

- characterized by deposits of white, fibrillar material on the anterior lens surface and throughout the anterior chamber
- is associated with a secondary open-angle glaucoma
- Vogt is credited, in the mid-1920s, with the original description of white flakes on the anterior surface of the lens being associated with glaucoma in some patients
- He referred to this as "senile exfoliation" of the lens.
- Dvorak-Theobald recommended the term "pseudoexfoliation" of the lens capsule when referring to this syndrome to separate it from true exfoliation.
- The association of pseudoexfoliation syndrome and glaucoma varies throughout the world.
- In Sweden and Norway, over 60% of open-angle glaucoma patients have pseudoexfoliation syndrome.
- In the United States, this association falls well below 15%.
- The prevalence of pseudoexfoliation syndrome (without glaucoma) varies widely among reports and in various geographic regions.
- The prevalence of pseudoexfoliation syndrome in the United States is between 1 and 5% of the population and increases with age.
- In Scandinavian countries, however, pseudoexfoliation syndrome is found in over 50% of the population.
- No clear hereditary patterns have been identified in pseudoexfoliation syndrome.
- Throughout the world, women are found to have a slightly higher prevalence of the syndrome, but the incidence of glaucoma in the syndrome shows no sex predilection.
- Classically, the material is deposited on the central portion of the anterior lens capsule in a circular pattern with a surrounding clear zone.
- With pupillary dilation, this clear zone can be identified, and a second zone of deposition is seen on the peripheral lens surface and anterior surface of the zonules.
- The material deposited on the central surface of the lens may have scroll-like edges and appear much the same as true exfoliation of the lens.
- The more peripheral deposits on the anterior lens assume the radial orientation of the underlying zonular attachments to the lens.
- White flecks of the pseudoexfoliation material may also be seen on the pupillary margin of the iris, as well as on the anterior surface of the iris.
- Peripupillary iris atrophy is often present with loss of pigmentation and transillumination defects.
- Similar deposits may be found on the endothelial surface of the cornea and in the anterior chamber angle.
- Hypoperfusion and neovascularization of the iris have been reported in the late stages of pseudoexfoliation syndrome.
- Iridodonesis and phacodonesis may also occur with this syndrome.
- Gonioscopic findings are noteworthy, not only for pseudoexfoliation deposits, but also for increased pigmentation of the angle structures.
- Pigmentation of the angle structures is usually patchy in pseudoexfoliation syndrome, as compared with the more uniform pigment distribution seen in pigmentary dispersion syndrome.
- In addition, a line of pigment deposition anterior to Schwalbe's line is often present in pseudoexfoliation syndrome (Sampaolelís' line).
- Decreased density of corneal endothelial cells may be seen by specular microscopy in this syndrome.
- Intraocular pressure elevation may occur months to years after the discovery of pseudoexfoliation syndrome.
- Intraocular pressures tend to be greater in the eye with the more significant deposition of pseudoexfoliation material when the deposition is asymmetric.
- Clinical findings of pseudoexfoliation syndrome may be asymmetric between the two eyes, and although unilateral cases of pseudoexfoliation have been reported, most cases are bilateral.
- The pseudoexfoliation material is a fibrillar protein with many characteristics of the connective and elastic tissues of basement membrane.
- Debate over the source of the pseudoexfoliation material continues.
- Some investigators think the material is derived from the lens epithelium, while others believe its source may be multifocal.
- The material has been found in the basement membranes of the ciliary body, iris, trabecular meshwork, conjunctival vessels, and iris vessels, as well as on the lens surface.
- Although the actual source of the exfoliation material is not known, it is generally accepted that elevation of intraocular pressure results from obstruction of the trabecular meshwork by the fibrillar material.
- Some studies have suggested that there is also an underlying baseline deficiency of aqueous humor outflow in these eyes.
- Other investigators believe that pseudoexfoliative glaucoma is an ocular manifestation of a systemic disease.
- The differential diagnosis of pseudoexfoliation syndrome includes true exfoliation of the lens and pigmentary glaucoma.

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<tr>
<th>DIFFERENTIAL DIAGNOSIS OF PSEUDOEXFOLIATIVE GLAUCOMA</th>
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<tbody>
<tr>
<td>Primary open-angle glaucoma</td>
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<td>Primary angle-closure glaucoma</td>
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<tr>
<td>Pigmentary glaucoma</td>
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<tr>
<td>Inflammatory glaucoma</td>
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<tr>
<td>True exfoliation/capsular delamination</td>
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• True exfoliation of the anterior capsule of the crystalline lens appears as white, scroll-like projections, similar in appearance to pseudoexfoliation and capsular separation from the underlying lens. Also, it is typically secondary to infrared radiation exposure (glass blower's cataracts).

• The management of glaucoma associated with pseudoexfoliation syndrome is generally the same as with any open-angle glaucoma.

• Some practitioners believe that medical therapy is more difficult in this syndrome.

• As with pigmentary dispersion syndrome, argon laser trabeculoplasty has been shown to be effective in these patients.

• Conventional filtration surgery also is effective.

• The long-term prognosis for control of the intraocular pressure is good in patients with pseudoexfoliation syndrome and glaucoma.

• Lens extraction is not necessary for the control of the glaucoma, as was once thought.

• Caution should be used when doing cataract extraction in patients with pseudoexfoliation syndrome.

• Up to a sevenfold increase in the incidence of vitreous loss has been noted during extracapsular cataract extraction in patients with pseudoexfoliation syndrome.

• Rupture of the posterior lens capsule also occurs more frequently in these patients.

• Zonular insufficiency and decreased capsular integrity are thought to contribute to these complications.

Patients should be examined, preoperatively, for evidence of zonular dialysis and phacodonesis.