Overview: Several different systemic diseases can present with ocular manifestations, which can include specific types of cataracts. This PowerPage will review the most common cataracts associated with systemic diseases.

**Diabetes Mellitus**

- Cataracts in patients with diabetes most commonly present as cortical or posterior subcapsular cataracts that occur at an earlier age in patients with poor blood sugar control as compared to age-matched controls.
- These cataracts often progress more rapidly and are more closely related to poor glucose control than duration of the disease.
- Classic diabetic cataracts occur due to a high level of glucose present in the aqueous humor, which diffuses into the lens. Within the lens, glucose is metabolized by aldose reductase into sorbitol, which then accumulates within the lens. This results in a subsequent osmotic over-hydration of the lens substance.
- In mild cases, this change may affect the refractive index of the lens, which can then lead to fluctuations in refraction related to changes in plasma glucose levels.
- Cortical fluid vacuoles can also develop in these patients, which can evolve into more dense opacities.
- Classic diabetic cataracts consist of snowflake cortical opacities that may either resolve spontaneously or mature rapidly within a short period of time.

**Myotonic Dystrophy**

- Approximately 90% of patients with myotonic dystrophy will develop cataracts in the third decade of life.
- Cataracts in these patients often present as central, polychromatic, iridescent, cortical crystals that have a “Christmas tree” appearance.
- These initially innocuous opacities typically evolve into a visually disabling stellate posterior subcapsular cataract by the fifth decade.
- In some cases, the presence of this type of cataract may precede myotonia symptoms.

**Wilson's Disease**

- Although a less frequent manifestation of Wilson’s disease, patients with this condition may present with the classic “sunflower cataract.”
- This type of cataract occurs due to fine copper deposition (also known as chalcosis lentis) beneath the anterior and posterior lens capsule that forms a disc-like opacity axially with radiating spoke- or petal-like extensions.
- On slit-lamp examination, the opacities may appear to be of various colors including reds, blues, greens, yellows, and browns.
**Hypocalcemia**

- Patients with hypocalcemia often develop cataracts that present as small white dot lens opacities that can eventually aggregate into larger flakes.
- The pathophysiology of this type of cataract is believed to be due to the fact that calcium is necessary to maintain membrane integrity, and that calcium deficiency will lead to membrane disruption and increased permeability.

**Atopic Dermatitis**

- Approximately 10% of patients with severe atopic dermatitis develop cataracts in the second to fourth decades of life.
- These lens opacities are often bilateral and present as dense shield-like anterior subcapsular plaques, which commonly cause wrinkling of the anterior capsule.
- Cataracts in these patients may progress very quickly.

**Neurofibromatosis-2 (NF-2)**

- Some patients with neurofibromatosis type 2 will develop cataracts that may occur in either one or both eyes, and often present in childhood.
- NF-2 is associated with posterior subcapsular or posterior cortical opacities.

**Fabry’s Disease**

- Ocular manifestations of Fabry’s disease are typically not visually significant; however, these are very important as they can be markers for the disease with diagnostic and prognostic implications.
- Ocular signs in patient’s with Fabry’s are a result of deficient activity of the lysosomal hydrolase, alpha-galactosidase A. This leads to deposition of glycosphingolipids in ocular structures, including the conjunctival vasculature, cornea (corneal verticillata), lens (nuclear sclerosis), and retinal vasculature.