The differential diagnosis of bilateral corneal crystals includes:

Corneal Stromal dystrophies
- Macular
- Granular
- Lattice
- Bietti's Crystalline dystrophy
- Schneider's crystalline dystrophy

Medication Toxicity- Topical Fluroquinolones

Corneal Infections- Streptococcus viridans

Systemic diseases
- Cystinosis
- Lymphoproliferative disorders – Multiple myeloma, Waldenstroms macroglobinemia, monoclonal gammopathy of unknown cause.

The three clinical forms of Cystinosis:

a) Nephropathic Infantile Cystinosis is the most frequent and the most severe form of the disease.[1] Clinical symptoms present in the first few months of life and are characterised by poor growth, polyuria, polydypsia, dehydration and acidosis. By the age of six months, full blown Fanconi syndrome is usually present and causes clinical symptoms of polyuria, polydypsia, failure to thrive, growth retardation, vomiting, periods of dehydration, constipation, developmental delay and rickets. [2] The Fanconi Syndrome involves failure of the renal tubules to reabsorb water, electrolytes, bicarbonates, phosphate, calcium, glucose, carnitine, and aminoacids. Excessive loss of calcium and phosphate can cause the development of nephrocalcinosis and the formation of renal stones. [2] Without therapy, cystine accumulation occurs in virtually all organs and tissues, including bone marrow, liver, intestine, muscle, brain, spleen, thyroid, pancreas and testes. [1] Affected individuals typically develop hypothyroidism at the end of the first decade of life. [1]

Photophobia develops when the cornea becomes packed with crystals, generally at the end of the first decade of life. Late ocular complications include: i) Anterior Segment Problems- Crystals in the anterior lens surface, band keratopathy, peripheral corneal neovascularisation and posterior synechiae, and ii) Posterior Segment problems- pigmentary retinopathy with degeneration of the photoreceptors that contributes to the impaired visual function in the late stage of the disease. Electroretinogram (ERG) is used to confirm retinopathy. [1]

b) Nephropathic Juvenile Form (Intermediate Cystinosis); renal and ocular involvement occurs later, during teenage or early adulthood.

c) Non Nephropathic adult form; Ocular non nephropathic cystinosis manifests only with complaints of photophobia due to cystine accumulation in the cornea. The kidney, retina and other organs are spared. [2]

Treatment for Cystinosis

a) Specific: Cysteamine, (1.3-1.9 g/m^2) in four daily doses helps reduce the accumulation of cystine in the cells, lowering the amount of damage done to tissues. (Accumulation of the amino acid cystine amplifies and alters apoptosis in such a way that cells die inappropriately). [2]

b) Systemic: As systemic treatment has no effect on corneal cystine crystals, topical cysteamine is indicated. Topical Cysteamine 0.5% given at least six times in a day is effective in dissolving the corneal crystal deposition and in ameliorating the photophobia. After penetrating keratoplasty, grafts usually maintain clear with minimal crystal deposition. [2]

c) Supportive: Correction of metabolic acidosis, oral phosphorous supplements, growth hormone therapy and thyroxin. Renal transplantation corrects renal failure but does not prevent progression of the disease in non renal organs.