

Review of Hereditary Nephropathies

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Hereditary nephropathies include disorders of the glomerular basement membranes due to variable mutations in the genes encoding the minor α chains of collagen IV. Others include various metabolic disorders due to enzyme deficiencies and lysosomal storage abnormalities leading to abnormal accumulations of glycosphingolipids and lipids in cells and tissues. Crystal accumulation in cystinosis and oxalosis are rare forms of hereditary nephropathy. Familial gout is another.

Fibronectin and collagen III glomerulopathy cause unusual deposition in glomeruli and there are also a number of other glomerulopathies such as FSGS and HUS which have hereditary variants. Sickle cell disease can lead to a nephropathy. Some forms of amyloid are hereditary.

Electron microscopy in many of the hereditary nephropathies is characteristic and crucial to the diagnosis in some. Diagnostic difficulties are often encountered however as some of the ultrastructural and other features overlap with appearances in other non hereditary nephropathies.

Reference: Heptinstall's Pathology of the Kidney 5th edn Vol 2 Section V Chapters 26-31