NEPHROTIC and NEPHRITIC SYNDROMES

	Disease	**Most Frequent Clinical Presentation**	**Pathogenesis**	**Light Microscope**	**F.M. (Fluorescence Microscope)** **E.M. (Electron Microscope)**	**Age Group Affected**	**Treatment and** **Outcome**
	Minimal Change Disease (Lipoid Nephrosis)	• Selective proteinuria (Albumin)	• Loss of foot processes • Loss of GBM polyanionic sites • Appearance of villi on epithelial cells	Normal Lipid in tubules	F.M. = negative E.M. = loss of foot processes, lipid vacuoles	#1 cause of Nephrotic Syndrome in children, esp. boys younger than 6 yrs. old.	Responds well to corticosteroids. <u>No</u> progression into chronic renal failure
	Focal Segmental Glomerular Sclerosis	• Non-selective proteinuria • Hypertension • Microscopic hematuria	• Idiopathic • Lower renal mass (in obese) • 2 causes: heroin use, HIV	• Focal and segmental sclerosis • Hyalinosis • Adhesions to Bowman's Capsule • Hypercellular mesangium • Thick B.M.	F.M. = IgM, C3 E.M. = Loss of foot processes, detachment of epithelium from B.M.	Majority occur in older children. Also occurs in adults.	Does not respond to corticosteroids. Leads to renal failure.
	Membranous Nephropathy (Glomerulonephritis)	• Persistent proteinuria	• Idiopathic • 2 causes: carcinomas, SLE, hepatitis, Diabetes Mellitus, thyroiditis, drugs.	• Glomeruli are enlarged yet normocellular • No cellular proliferation • B.M. Thickening	F.M. = "Spike and Dome." Granular IgG, C3 E.M. = Subepithelial immune deposits in B.M., thickened B.M.	#1 cause of Nephrotic syndrome in adults	Benefit of corticosteroids is unknown.

Diabetic Nephropathy **(Diabetic Glomerulosclerosis)**	• Proteinuria • No hematuria	• Diabetic microangiopathy • Thickened B.M. • Massive mesangial growth • "Kimmelstiel Wilson" nodular glomerulosclerosis • Diffuse glomerulosclerosis	• Kimmelstiel-Wilson Nodules are pathognomonic. • Massive mesangial hypercellularity.	F.M. = negative E.M. = massive mesangial growth, thickened B.M.	Diabetics	Progresses to renal failure	
Reoidosisnal Amyl		Subendothelial and mesangial amyloid deposits	• Amyloid deposits are initially mesangial, producing mesangial widening without hypercellularity. • Later, the amyloid obliterates the lumen • PAS(-) • Congo-Red (+)	F.M. = negative E.M. = characteristic criss-cross fibrillary proteins.	Any age group	Severe amyloid infiltration leads to renal failure	
	Alport Syndrome (Hereditary Nephritis)	• Recurrent hematuria before age 20 • Hypertension • Deafness and ocular problems	Structural defect in Collagen IV leads to leaky basement membranes.	Looks normal	F.M. = negative E.M. = glomerular B.M. splitting	Symptoms appear before age 20	Progresses to renal failure
	Benign Familial Hematuria (Thin B.M. Disease)	• Recurrent hematuria • Most frequent cause of asymptomatic hematuria.	Reduced thickness of glomerular B.M.	Looks normal	F.M. = negative E.M. = reduced glomerular B.M. thickness		

	Acute (Post-Streptococcal) Glomerulonephritis	• Acute nephritis • Abrupt oliguria, hematuria, facial edema, hypertension.	• Immune-complex mediated (Type-III hypersensitivity) • Occurs after Streptococcal pharyngitis or Hepatitis-B • High ASO-titer, low C3	• Glomerular hypercellularity • Increase in endothelial cells, mesangial cells, and PMN's. • No increase in epithelial cells. • No B.M. thickening	F.M. = "lumpy-bumpy" granular deposits of IgG and C3 E.M. = Subepithelial (not subendothelial) "humps," otherwise normal appearing B.M.	Common renal disease in childhood	Return to normal in 8 weeks. Complete recovery without treatment (especially in kids) within 3 years.
	SLE Nephropathy	Degree of kidney involvement correlates with prognosis in SLE.	Anti ds-DNA antibodies.	• WHO I: Normal • WHO II: Increased mesangial matrix • WHO III: Focal proliferation • WHO IV: Diffuse proliferation, worst. • WHO V: Identical to Membranous Nephropathy	F.M. = IgM, IgG + C3 • Type-I: Granular appearance • Type-II: Pseudo-linear appearance		
	Focal Segmental Glomerulonephritis	IgA Nephropathy (Berger's Disease): Most common primary glomerulonephritis	Circulating IgA + fibronectin (due to chronic liver disease)	• Mesangial cell proliferation	F.M. = Granular appearance, IgG + C3 E.M. = Mesangial deposits	Young men 15-30	

Henoch-Schonlein Purpura		Same as above, plus systemic disease: purpura of extremities, arthritis, colicky abdominal pain.	• Mesangial cell proliferation, more serious than above.	F.M. = Granular appearance, IgG + C3 E.M. = Mesangial deposits	Children		
Endocarditis	S. Aureus	• Subepithelial immune deposits	F.M. = Granular appearance, IgG + C3		Kidney disease resolves when infection is cured.		
Rapidly Progressive Crescentic Glomerulonephritis	• Wegener's: kidney + upper respiratory tract. • Anuria • Oliguria	• Inflamed glomerular capillaries • ANCA (+)	• Cells accumulate in Bowman's Capsule • Fibrin trapped in glomeruli • Epithelial cell proliferation • Macrophage, PMN infiltrates	F.M. = Pauci-immune. Irregular E.M. = wrinkling, discontinuity of B.M.		Must be treated or it will go to renal failure within weeks.	
Goodpasture Syndrome **(Anti-BM Antibody Disease)**	Lung (hemoptysis) + kidneys (hematuria)	Anti-B.M. antibodies, against Type-IV collagen	Similar to Crescentic glomerulonephritis, as above.	F.M. = Linear pattern, IgG + C3 E.M. = No immune complex deposits	Males 25-30	Responds to immunosuppressive therapy and plasmapheresis	
Membranoproliferative Glomerulonephritis **(Mesangiocapillary Glomerulonephritis)**			• B.M. thickening and cellular proliferation • Mesangial expansion makes glomerular B.M. appear as though it were in two layers	E.M. = "Tram-track" appearance, resulting from double-layer appearance of glomerular B.M.			