Glomerulonephritis, X-Linked Mesangiocapillary

**Alternative Names**
Mesangiocapillary Glomerulonephritis, X-Linked
MCGNX
Membranoproliferative Glomerulonephritis

**Record Category**
Disease phenotype

**WHO-ICD**
Diseases of the genitourinary system > Glomerular diseases

**Incidence per 100,000 Live Births**
6-10

**OMIM Number**
305800

**Mode of Inheritance**
? X-linked predisposition

**Gene Map Locus**
N/A

**Description**
X-linked mesangiocapillary glomerulonephritis is a kidney disorder causing decreased kidney function because of inflammation and changes in the tissues of the internal kidney structures. It is histologically characterized by intense glomerular hypercellularity, mainly due to mesangial proliferation, and diffuse thickening of the glomerular basement membrane with the appearance of ‘double contours’. Two distinct histological forms have been identified – Type I mesangiocapillary glomerulonephritis and Type II mesangiocapillary glomerulonephritis. Type I mesangiocapillary glomerulonephritis is characterized by massive mesangial proliferation, mesangial matrix expansion and diffuse thickening of the glomerular basement membrane, while Type II mesangiocapillary glomerulonephritis presents dense homogenous deposition along the glomerular basement membrane and in the mesangium. A third variety in which there are not only sub-endothelial deposits but also numerous sub-epithelial and intramembranous deposits, associated with replication of the lamina densa and frequently disruption of the whole basement membrane.

**Molecular Genetics**
N/A

**Epidemiology in the Arab World**

**Bahrain**
Al Arrayed et al. (2004) conducted a retrospective study and analyzed 498 renal biopsies of Arab and non-Arab patients with proteinuria, hematuria, and mild to moderate renal impairment during a period of 13 years (between January 1990 and December 2002). Among the 498 samples, 375 (75.3%) belonged to Arab patients originating from Bahrain, United Arab Emirates, Oman, Yemen, Saudi Arabia, Kuwait, Qatar, North and East Africa, and the Mediterranean region. The male:female ratio was 1.4:1. Among the Arab cases, glomerular disease constituted 66.7% of their total. Primary glomerular diseases constituted 63.6% of glomerular lesions; secondary glomerular diseases, 36.4%. Among the primary glomerular diseases, minimal change disease was the commonest lesion (27.7%). The second commonest diagnosis was focal segmental glomerulosclerosis (22.6%). Lupus nephritis (41.8%) formed the commonest cause of secondary glomerular diseases followed by diabetic nephropathy (33%) and hypertensive nephropathy (17.6%). Al Arrayed et al. (2004) found out that glomerular diseases were more common in males than in females and calculated their incidence rate in Bahrain to be 5.8/100,000 per year. This is a much higher incidence than the one recorded in the United Arab Emirates [See also: United Arab Emirates > Yahya et al. (1998)]. Al Arrayed et al. (2004) suggested that this variation may be due to the time span of the reviews and/or because of the various biopsy policies adopted by different centers.
Kuwait
Zaki et al. (1989) studied 55 Arab children with primary nephrotic syndrome over a 5-year period. There were 35 boys and 20 girls with a mean age of 5.3 years. The annual incidence was 7.2 and 6.0 per 100,000 children below 10 and 12 years of age, respectively. An initial response to steroids was noted in 84% with almost 50% responding within 1 week of therapy. Nine patients did not respond to steroids; histopathological classification of their renal biopsies showed 5 cases of membranoproliferative nephritis. Microscopic haematuria was noted at presentation in 7 of 46 steroid responders, in all 5 patients with membranoproliferative disease and in 1 of 3 with focal segmental glomerulosclerosis. [See also: Bahrain > Al Arrayed et al. (2004)].

Lebanon
Mourani et al. (1998) conducted repeated percutaneous biopsies in allograft kidneys and in native kidneys in 61 patients from Lebanon using small disposable needles in a well sedated child. No major complications were noted. Mourani et al. (1998) indicated that mesangiocapillary glomerulonephritis and focal segmental glomerulosclerosis were the predominant histological findings in patients from Lebanon and called for data to be collected from other centers to evaluate the real incidence of the different entities of renal diseases in Lebanon.

Oman
[See also: Bahrain > Al Arrayed et al. (2004)].

Qatar
[See also: Bahrain > Al Arrayed et al. (2004)].

Saudi Arabia
Abdurrahman et al. (1989) evaluated 16 Saudi children with early onset nephrotic syndrome over a 5-year period. This sample represented 17% of the 92 cases of childhood nephrotic syndrome seen during the period. Onset of the nephrotic syndrome was less than or equal to 3 months of age in four patients. Ten of the patients developed renal failure. Eight patients died, seven of them by 1 year of age. Two patients who were given renal transplants had functioning grafts without recurrence of the disease. Renal biopsy in two patients showed mesangio proliferative glomerulonephritis. In 1990, Abdurrahman et al. described the clinicopathological features in 119 Arab children from Saudi Arabia with the nephrotic syndrome. The clinical and laboratory data were similar to those described in other parts of the world. However, mesangial proliferative glomerulonephritis (MesPGN) was found in 21 of 66 biopsies (31.8%), giving a frequency of 17.6% of all children with the nephrotic syndrome. Onset of the nephrotic syndrome was at less than 1 year of age in 17 patients (14.3%). There were nine deaths, all in patients with end-stage renal disease: six of the deaths occurred in infants. Abdurrahman et al. (1990) concluded that the pattern of childhood nephrotic syndrome in Saudi Arabia is different from that in tropical countries.

In 1996, Mitwalli et al. reviewed the clinical data and renal biopsies of 186 adult patients found to have nephropathy and seen at the Security Forces Hospital, Riyadh, over a 5-year period (1989 to 1994). Mitwalli et al. (1996) found out that Mesangio proliferative glomerulonephritis was the second most common lesion (21.1%), following primary glomerular disease. This was followed by membranous glomerulonephritis (13.6%), immunoglobulin A nephropathy (IgAN) (13.6%), membranoproliferative glomerulonephritis (9.5%), and minimal change disease (1.4%).

In 2003, Kari reported the spectrum of membranoproliferative glomerulonephritis in eight Arab patients from Saudi Arabia with membranoproliferative glomerulonephritis type I and type II. The mean age of the patients at presentation was 2.4 +/- 1.2 years. All patients presented with a steroid resistant nephrotic syndrome. None had macroscopic hematuria. However 5 (62.5%) were hypertensive at presentation. The mean follow-up between presentation and last visit was 1.1 +/- 0.7 years; range 0.1-2. Three patients were siblings and their parents were second-degree cousins. Another patient had a brother who had a renal failure following steroid resistant nephrotic syndrome (SRNS), but the histological cause of his SRNS was not known. Kari (2003) concluded that membranoproliferative glomerulonephritis seems to present at earlier age in Arab children and tends to have a severe course with rapid progression to end stage renal disease.

[See also: Bahrain > Al Arrayed et al. (2004)].

United Arab Emirates
Abou-Chaaban et al. (1997) studied the pattern of pediatric renal diseases among children in the Dubai Emirate during the period from 1991 to 1996. In this period, a total of 712 pediatric patients, including 230 nationals of the United Arab Emirates, were seen with various renal problems. A total of 50 patients
presented with chronic renal failure, of which seven had glomerulonephritis.

There is little data on the spectrum of renal diseases in the United Arab Emirates. A renal diseases registry has been set up in an attempt to address this issue nationwide. In 1998, Yahya et al. reported the first outcome of this endeavor, a retrospective histopathologic analysis of 490 native kidney biopsies performed on adult patients presenting to four hospitals in the Emirate of Abu Dhabi from 1978 to June 1996. Primary glomerular disease accounted for 77.1% of all biopsies. Chronic proliferative glomerulonephritis as a group was the predominant pathology (36.2%), followed by idiopathic membranous glomerulopathy (20.1%), focal segmental glomerulosclerosis (18.3%), minimal change nephropathy (18.3%), and IgA nephropathy (6.3%). Of the patients with secondary kidney diseases, 33 (40.7%) had systemic lupus erythematosis, 27 (33.3%) amyloidosis, 14 interstitial nephropathy, and seven diabetic nephropathy. Yahya et al. (1998) also analyzed kidney biopsies of 187 patients with primary glomerular disease who presented with the nephrotic syndrome. In this group idiopathic membranous glomerulopathy, proliferative glomerulonephritis, and minimal change glomerulopathy was found in almost equal proportions (28.3%, 26.6%, 26.2%) with focal segmental glomerulosclerosis (15.4%) accounting for the bulk of the remainder.

[See also: Bahrain > Al Arrayed et al. (2004)].

Yemen
[See also: Bahrain > Al Arrayed et al. (2004)].

References


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