

Paediatric Nephrology at the University Hospital of the West Indies

A Walk through Time

M Miller

ABSTRACT

Paediatric nephrology at the University Hospital of the West Indies has grown over the last five decades into an established paediatric subspecialty offering to Jamaica and other Caribbean territories the benefit of paediatricians with training and exposure in this field. Dissemination of information to medical practitioners within the island has reduced mortality and morbidity associated with potentially treatable atypical renal disease. Clear investigative guidelines for urinary tract infection have made earlier diagnosis of urological malformations possible. Patterns of glomerular disease in Jamaican children have now been aetiologically and histologically documented. Children with chronic kidney disease now have clear management guidelines and the possibility of renal replacement therapy. Future goals include community education about renal disease and the development of a paediatric dialysis and transplantation unit.

Keywords: Glomerulonephritis, nephrotic syndrome, renal failure

Nefrología Pediátrica en el Hospital Universitario de West Indies

Un Recorrido a Través del Tiempo

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RESUMEN

La nefrología pediátrica en el Hospital Universitario de West Indies se ha desarrollado en las últimas cinco décadas, convirtiéndose en una sub-especialidad pediátrica establecida, que ofrece a Jamaica y otros territorios caribeños, el beneficio de pediatras con entrenamiento y experiencia en este campo. La diseminación de la información entre los médicos dentro de la isla, ha reducido la mortalidad y la morbilidad asociadas con la enfermedad renal atípica potencialmente tratable. Los claros lineamientos de la investigación de las infecciones del tracto urinario, han hecho posible un diagnóstico más temprano de las malformaciones urológicas. Los patrones de enfermedad glomerular en los niños jamaicanos han sido ahora documentados tanto etiológica como histológicamente. Los niños con enfermedad renal crónica tienen a hora a su disposición lineamientos claros para el tratamiento así como la posibilidad de la terapia del reemplazo renal. Las metas futuras incluyen la educación de la comunidad sobre la enfermedad renal y el desarrollo de una unidad pediátrica de diálisis y trasplante.

Palabras claves: Glomerulonefritis, síndrome nefrótico, insuficiencia renal

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INTRODUCTION

Paediatric nephrology as a subspecialty did not formally begin at the University Hospital of the West Indies (UHWI)

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until December 1984 with the arrival of Dr Maolynne Miller, the first paediatric nephrologist. However the first paediatrician with interest in this field was Dr Dawn Swaby.

Glomerulonephritis

It was a common practice of physicians then, in some hospitals, to deem all types of acute glomerulonephritis, post-streptococcal (PSGN), and expect a good prognosis in spite of atypical features. Many of these atypical children did not

undergo renal biopsy and progressed to chronic renal failure (1). The initial thrust of paediatric nephrology at UHWI was education at all levels – medical students, residents, consultants, paediatricians and physicians in general practice – to recognize presentations of renal disease which required urgent nephrological referral. This information was spread through islandwide meetings with various medical groups. Residents in training were able to take back to their territory (if non-Jamaican) and to their practices, information on paediatric nephrology. To facilitate this, two paediatric nephrology manuals were written – in 1988 (2) and a revision in 2002 (3) with paediatric norms, investigative and management protocols.

Renal Failure

Prior to 1984/85, the management of chronic renal failure in children was not standardized and the absence of 1-hydroxylated 25-OH Vitamin D resulted in rampant rachitic deformities. Dialysis for acute renal failure was generally unavailable and pulmonary oedema from renal failure was treated with mechanical ventilation. There were no facilities for chronic dialysis or transplantation. After December 1984, acute peritoneal dialysis became possible and paediatric renal biopsies could be performed with greater ease. As the association with the adult nephrology team evolved into the dynamic unit it now is, children were able to benefit from haemo- and peritoneal dialysis. In fact, a nine-year old actually received a renal transplant in 2001 which served him well. We are now able to offer chronic automated peritoneal dialysis and haemodialysis to children weighing as little as 13 kilograms.

Urinary Tract Infection

Prior to December 1984, children with urinary tract infection (UTI) were investigated only if infections were recurrent, associated with a clinically obvious urological abnormality, or if the child was male. Children with urological pathology were often diagnosed late, already with chronic kidney disease (1). Since then, local guidelines were formalized and disseminated to ensure adequate investigation of Jamaican children (4, 5).

In a 1984–2005 retrospective study of 301 children with UTI in a combined general paediatric and paediatric nephrology referral service, renal ultrasound was normal in 88% with vesico-ureteric reflux and 75% of those with posterior urethral valves, making it an inadequate screening tool for lower tract abnormalities (6).

Prospective research has documented that *Escherichia coli* is the commonest uropathogen in Jamaican children < age 12 years with first UTI (7). *Escherichia coli* is highly resistant to ampicillin which is thus an inappropriate first line treatment for UTI. Overall, the greatest sensitivity was to gentamycin followed by amoxil clavulanate and cotrimoxazole. It now seems reasonable to use cotrimoxazole as first line treatment in children in the 1 to under 12-year age group

in whom the resistance pattern is similar to amoxil clavulanate.

Nephrotic Syndrome

In the early days, outpatient urine protein testing was not done because of the prohibitive cost of commercial Labstix™. Children with nephrotic syndrome frequently needed admission as oedema was the only indicator of relapse. With the use of home testing by test tube using sulphosalicylic acid, relapses are diagnosed earlier and admissions for nephrotic syndrome have drastically reduced.

Complete serological evaluation of all children with nephrotic syndrome is now practised resulting in improved ability to make aetiological diagnoses. Hepatitis B infection was found to be the cause of nephrotic syndrome in 6% of otherwise asymptomatic children with nephrotic syndrome and the commonest cause of childhood membranous nephropathy (8). These data were able to support the appeal for Hepatitis B immunization in Jamaican children, and since the implementation of vaccination, no new cases have been observed. The first published cases of HTLV-1 associated renal disease were Jamaican children with infective dermatitis (9) resulting in the inclusion of this investigation in children with renal disease.

Patterns of Renal Disease

Glomerulonephritis

Over the years, the pattern of renal disease has evolved. In a retrospective review of glomerulonephritis in Jamaican children from 1978–1982, 95% of cases were postinfectious with PSGN accounting for 98% of infections. The only two renal biopsies performed revealed mesangial proliferative glomerulonephritis (MesPGN) with focal sclerosis and diffuse proliferative glomerulonephritis (DPGN), respectively (10).

A review of renal biopsies in all Jamaican children less than 12 years of age was conducted between 1985–2008 to obtain information on the pattern of renal disease seen locally (11). Children were biopsied if they had nephrotic syndrome atypical for minimal change disease (MCD), unexplained or atypical glomerulonephritis, unexplained haematuria, proteinuria, or to stage renal involvement in systemic lupus erythematosus. Of the 270 children biopsied, the commonest histology was DPGN (27.7%), followed by MesPGN (25.5%).

Glomerular disease was associated with infection in fewer cases in the current study (32.4% vs 95%) with PSGN accounting for 75% of these compared with 98% in the earlier review. During the 24-year period, PSGN peaked between 1985 and 1988 and again 1993–1995 with corresponding trends in DPGN and MesPGN both of which histological appearances may be seen in PSGN.

In sickle cell (SS) disease, DPGN was again the commonest histology (47.4%), followed by MesPGN and membranoproliferative glomerulonephritis. There was no statistically significant difference between the frequency of

DPGN, PSGN and MesPGN in the children with SS disease and the remaining children, suggesting that children with SS disease have the childhood nephropathies endemic to Jamaica rather than renal disease due to the sickling process (12).

Nephrotic Syndrome

Prior to 1984, children less than 12 years of age with nephrotic syndrome most commonly had MCD followed by MesPGN (15%) and DPGN (7%). Most children did not have a renal biopsy (13).

Between 1984–1987, 26/27 Jamaican children presenting with nephrotic syndrome had features atypical for MCD and were biopsied. The commonest histology overall was MesPGN (30%) followed by MCD (26%) and focal segmental glomerulosclerosis (FSGS) [18%]. Diffuse proliferative glomerulonephritis accounted for only 4%. Nephrotic syndrome was idiopathic in 41% and postinfectious in the majority (59%) with PSGN being the commonest infection (81%). In unreferred patients, MCD was still the most common histology/presentation (45%) while in those referred for nephrological consultation, the frequency of MCD was 13% and MesPGN 44% (14).

In a histological review of all atypical cases of nephrotic syndrome in the island from 1985–2008, nephrotic syndrome was idiopathic in 63.6% and postinfectious in 24% (PSGN in 67%). Overall, MesPGN was the commonest appearance (31.2%) followed by MCD (22.9%) and DPGN (16.6%). In idiopathic nephrotic syndrome, MesPGN was again the most frequent histological appearance (43%) followed by MCD (29%) and FSGS and DPGN (10% respectively). The rise in frequency of FSGS observed with time in the international literature was not evident in Jamaican children (15).

Chronic Renal Failure

In the 16-year period between 1985 and December 2000, the annual incidence of chronic renal failure (CRF) in Jamaican children less than 12 years old was 3.2/million age-related population (34 children). Glomerular disease was the leading cause of kidney failure followed by obstructive uropathy, reflux nephropathy and renal dysplasia (41%). Postinfectious glomerulonephritis accounted for 26.5% of CRF and PSGN for 11.8%. Half of the children were already in chronic renal failure at the time of first presentation with renal disease and the mortality was 65%. Only three children (0.8%) had access to chronic dialysis locally (1).

In the six years that followed (2001–2006) the annual incidence of CRF had risen to 4.61/million age-related population [< 12 years] (18 children). Glomerular disease accounted for only 33.3% of CRF. Postinfectious glomerulonephritis had fallen to 16.7% and comprised only HIV associated nephropathy. There were no cases of PSGN leading to CRF. Congenital urological disease was the predominant cause of CRF (44.5%). Although these abnormalities were

being diagnosed and treated earlier, renal failure persisted in some. It appeared that education and early referral had reduced mortality from atypical PSGN. However, with the longer survival of HIV infected children, progressive renal disease was now apparent, usually associated with treatment noncompliance or failure. As before, about half of the children were already in CRF at first presentation with renal disease. Mortality had fallen to 44.4%. At the UHWI, 36% of children under 12 years old accessed chronic haemodialysis. There was one renal transplant in this group (16).

Plans for the Future

It seems that physicians now recognize and refer atypical cases earlier with improvement in outcome. The next goal is to take basic information about kidney disease to the community to ensure prompt presentation for medical care. The paediatric nephrology service will be strengthened in 2012 when a second paediatric nephrologist joins the University Hospital of the West Indies. Renal transplantation and a separate paediatric dialysis unit are our aspirations.

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