Congenital nephrotic syndrome and nonsteroidal anti-inflammatory drugs

Sir,

A female baby was born at 39 weeks gestational age after a smooth course of pregnancy. General edema started at 3 months of age and the diagnosis of congenital nephrotic syndrome (CNS) was considered based on heavy proteinuria (15 g/l), total serum protein (TSP) of 3 g/dl, and albumin of 10 g/dl. Kidney function was normal with a creatinine of (0.3 mg/dl). Secondary causes (syphilis, rubella, toxoplasmosis, cytomegalovirus, and hepatitis B) were excluded. C3 complement was decreased whereas C4 was normal, antinuclear antibodies were negative. Kidney biopsy showed several glomeruli which were shrunken, sclerotic, and covered by a corona of hypertrophic podocytes. Tubules were slightly dilated with focal cyst formation. No immune deposition was found. The diagnosis was compatible with diffuse mesangial sclerosis.The infant was treated with albumin 20% infusions, 3g/kg/d devided in three doses and progressively progressively increased to 4 g/kg/day. At the second week of admission patient was started on NSAID (Ibuprofen) with a dosage of 3 mg/kg/twice per day. Two days later adding ACEi (enalapril) was considered at 2 mg/kg/day once. Ten days later, we noted a decrease in the need for albumin infusion from and 2 weeks later we were able to withhold it. Kidney function remained stable with a serum creatinine of 0.4 mg/dl. Follow up 4 months later showed an infant with catch up growth and no edema with a total serum protein of 6.5 g/dl, and an albumin of 4.4 g/dl.

Congenital nephrotic syndrome is a rare kidney disorder characterized by heavy proteinuria, hypoproteinemia, and edema starting soon after birth, mostly within the first three month of life. Most are caused by genetic mutations which explain the resistance to steroids use. However, secondary causes have been identified and are treated according to the causes.[1] In contrast to most cases of idiopathic nephrotic syndromes, the CNS is resistant to immunosuppressive treatment, thus control of edema and prevention of complications is the aim of treatment. Some centers perform unilateral nephrectomy to reduce protein losses[2] with or without the usage of other medications like angiotensin converting enzymes inhibitors (ACEi) and nonsteroidal anti-inflammatory drugs (NSAID) such as indomethacin.

These drugs decrease proteinuria and consequently the need for albumin infusion. NSAID effect on decreasing proteinuria exceeds a simple decrease in glomerular filtration rate but seems to directly modify renal hemodynamics.[3] ACEi interfere with the renin-angiotenisn-aldosteron cascade and reduce proteinuria both in animals and humans.[4] The use of ACEi together with NSAID have been described to decrease but not a complete disappearance of proteinuria as noted here.

B. Aoun[1,2], C. P. Schmitt[3], T. Ulinski[4], H. Fakhoury[1], G. Kalkas[1], S. Sanjad[1,2]

[1]Department of Pediatric Nephrology, Rafic Hariri University Hospital, Beirut, Lebanon, [2]Faculty of Medicine, American University of Beirut, [3]Center for Pediatric and Adolescent Medicine, University of Heidelberg, Germany, [4]Armand Trousseau University Hospital University, Paris, France

Address for correspondence:
Dr. Bilal Aoun,
Department of Pediatric Nephrology, Rafic Hariri University Hospital, Beirut, Lebanon.
E-mail: drbilal77@hotmail.com

References

1. Tudorache E, Hogan J, Dourthe ME, Quinet B, Grimprel E, Sellier-Leclerc AL, *et al.* Congenital nephrotic syndrome with acute renal failure. Pediatr Nephrol 2012;27:47,49-50.
2. Jackson LW. Congenital nephrotic syndrome. Neonatal Netw 2007;26:47-55.
3. Vriesendorp R, Donker AJ, de Zeeuw D, de Jong PE, van der Hem GK, Brentjens JR. Effects of nonsteroidal anti-inflammatory drugs on proteinuria. Am J Med 1986;25;81:84-94.
4. Sreedharan R, Bockenhauer D. Congenital nephrotic syndrome responsive to angiotensin-converting enzyme inhibition. Pediatr Nephrol 2005;20:1340-2.

Access this article online	
Quick Response Code:	Website: www.indianjnephrol.org
	DOI: 10.4103/0971-4065.97149