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Heavy proteinuria in a child with single ventricle

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Backgrounds: It is well established that the patients who has heart disease can often result in secondary dysfunction or injury to their kidney. The proven factors that can cause glomerular damage from congenital heart disease are hypoxia, hyperperfusion, increased hydrostatic pressure and neurohormonal abnormalities.

Case: A 6-year-old girl admitted to our hospital with proteinuria which was persisted over 8 months. She was born with functional single ventricle and had Fontan's procedure within first year of her life. Kidney sonography showed diffuse increased echogenicity of both renal parenchyma and right kidney size was 7.3cm which was normal, but left kidney was 5.3cm. Laboratory test showed serum Hb 19.4 g/dL; Hct 56.0 %; T.Protein 7.8 g/dL; Albumin 4.1 g/dl; T.Cholesterol 202 mg/dL; BUN 20.0 mg/dL; Cr 0.67 mg/dL; eGFR 65.34 mL/min/1.73m². Urinalysis revealed protein 3+ and blood 1+. Urine protein/urine creatinine ratio was 6359.6 mg/g. Twenty four-hour urine protein was 1062.1 mg/day, which was nephrotic range. Renal biopsy showed that 14% of glomeruli had global sclerosis and remaining glomeruli was markedly increased in its size and hypercellular involving mesangial cells. No evidence of immune complex or autoantibody deposition in immunofluorescence. Glomerular basement membrane was diffusely thickened to a severe degree. There were localized subendothelial electron dense materials with mesangial interposition. Epithelial cell foot processes showed focal moderate effacement. Tubules showed loss with interstitial foam cells and fibrosis. Final diagnosis was glomerular enlargement associated with cyanotic heart disease. She received angiotensin-converting-enzyme inhibitor (0.2mg/kg/day) and continues to visit our clinic. After 2 months of medication, proteinuria decreased as 1+ on dipstick at her latest urinalysis.

Conclusions: Cyanotic heart disease can develop a nephropathy in children. Proper evaluation and management should be followed to prevent the patients from decreasing their renal function.

Keywords : preteinuria, cyanotic congenital heart disease, nephropathy, functional single ventricle