Glomerulomegaly, or glomerular enlargement, was first reported in patients with congenital cyanotic heart disease in 1953. This finding was initially noted on visual inspection and subsequently confirmed via morphometric measurements. Beyond cyanotic heart disease, several other conditions are also associated with glomerulomegaly, including cor pulmonale, pulmonary hypertension, polycythemia vera, sickle cell disease, obesity, alcoholism, fatty liver, and cystic fibrosis. More broadly speaking, glomerular hypertrophy is generally observed in both congenital (oligomeganephronia, congenital solitary kidney) and acquired (chronic kidney disease, post-nephrectomy) conditions associated with reduced nephron mass and is thought to be due to compensatory glomerular hyperfiltration that is required to maintain overall renal clearance.

The following are 10 points to remember about glomerulomegaly and the kidney in congenital cyanotic heart disease.

- Although glomerulomegaly used to be thought of as a benign condition, glomerular enlargement is now known to be associated with increased risk of glomerulosclerosis, progressive decline in kidney function, and poorer prognosis.
- In kidney transplant recipients, increasing glomerular size in the donor kidney is associated with a higher risk of late allograft dysfunction.
- A threshold glomerular size that predisposes to glomerulosclerosis has not yet been established.
- The putative mechanisms for the development of glomerulomegaly vary according to underlying physiologic abnormalities; these include increased right ventricular pressure causing congestion, increased blood volume, hypoxemia, hyperviscosity, and lipid abnormalities.
- In congenital heart disease, the term "cyanotic nephropathy" was coined because glomerulomegaly was thought to lead to a decline in kidney function. Proteinuria is the most frequently observed clinical abnormality.
- Glomerulomegaly in minimal change disease predicts subsequent progression to focal segmental glomerular sclerosis (FSGS).
- Glomerulomegaly in proliferative (class III or IV) lupus nephritis has been reported to be associated with lower probability of disease remission at 3 years post-induction therapy.
- Obesity-related glomerulopathy, defined as proteinuric renal disease in patients with a body mass index > 30 kg/m², is associated with glomerulomegaly and secondary FSGS. The classic presentation is moderate-to-massive proteinuria with a normal serum albumin level (in contrast to hypoalbuminemia seen in primary FSGS).
- Obesity is an independent risk factor for the development and progression of chronic kidney disease. Even in obese patients who have normal kidney function, body mass index positively correlates with glomerulomegaly. Interventions to reduce weight—including lifestyle modifications and, when unsuccessful, bariatric surgery—should be considered early for these patients.
- Blockade of the renin-angiotensin system is the mainstay of therapy to reduce glomerular hyperfiltration. The Eighth Joint National Committee on Prevention, Detection, Evaluation, and Treatment of High Blood Pressure (JNC-8) guidelines recommend angiotensin-converting enzyme inhibitors or angiotensin receptor blockers as first-line therapy for hypertensive patients with chronic kidney disease.

REFERENCES


