1995-2014

<25 patients

2015-2020

>25 patients

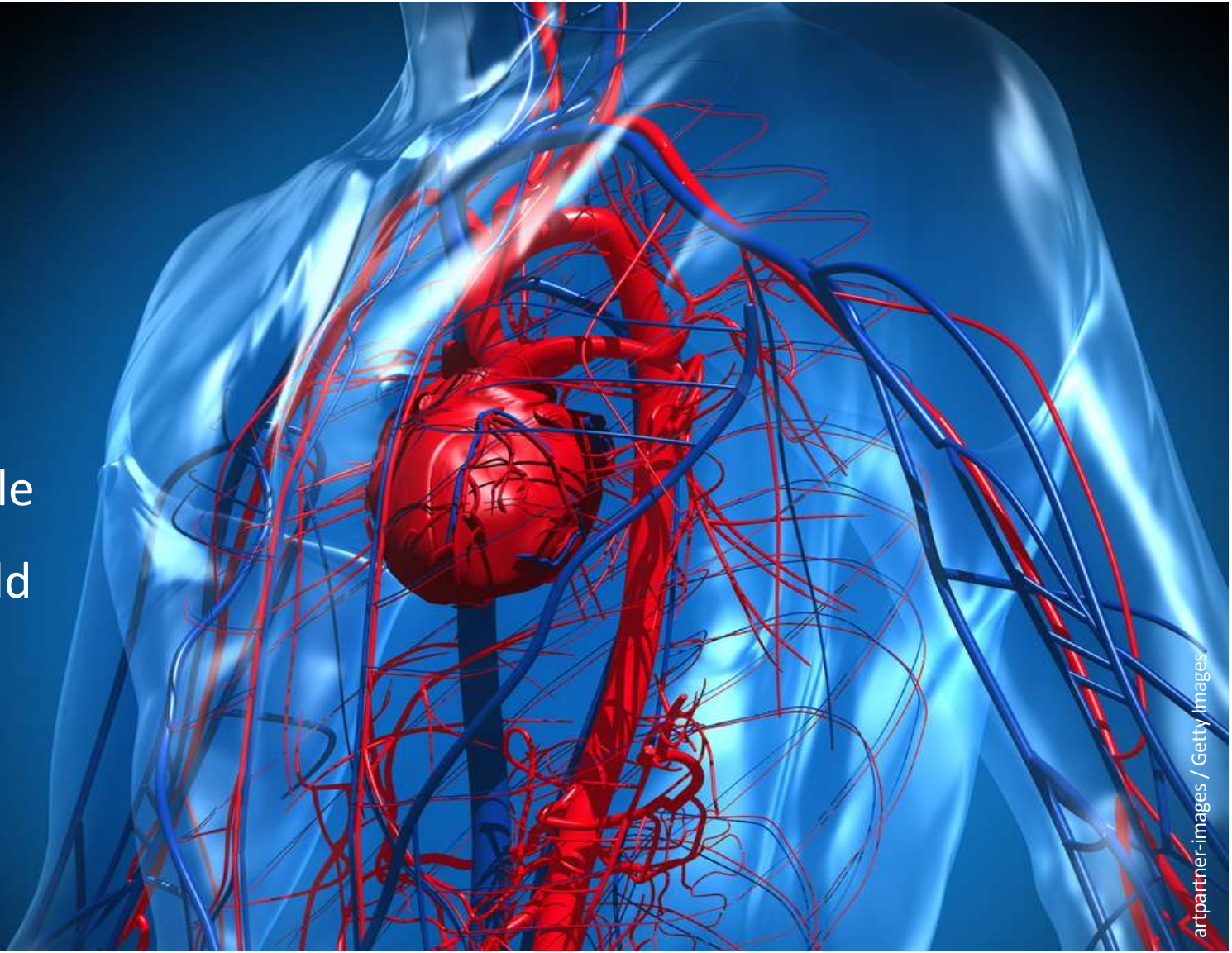
2021-2024

>50 patients

year	# patients	# sites and location
2009	**65**	multi, N. America
2014	**40**	single, Canada
2015	66	multi, France
2016	**293**	multi (45), N. America
2017	**105**	multi (22), N. America
2017	22	single, USA
2019	39	multi (11), Türkiye
2019	49	multi (1,700), Japan
2021	**85**	multi, Canada/Italy
2021	34	single, China
2022	48	single, China
2022	**131**	multi, N. America
2022	61	single, China
2023	46	multi (14), France
2023	337	multi, 41 countries
2024	17	single, USA
2024	**398**	multi, N. America
2024	179	multi (17), China
2025	**145**	multi, N. America

We have described ~1/3 of the ~1,500 cases of childhood chronic, systemic vasculitis reported globally

If all your blood vessels were arranged in a single line, how far would they stretch?



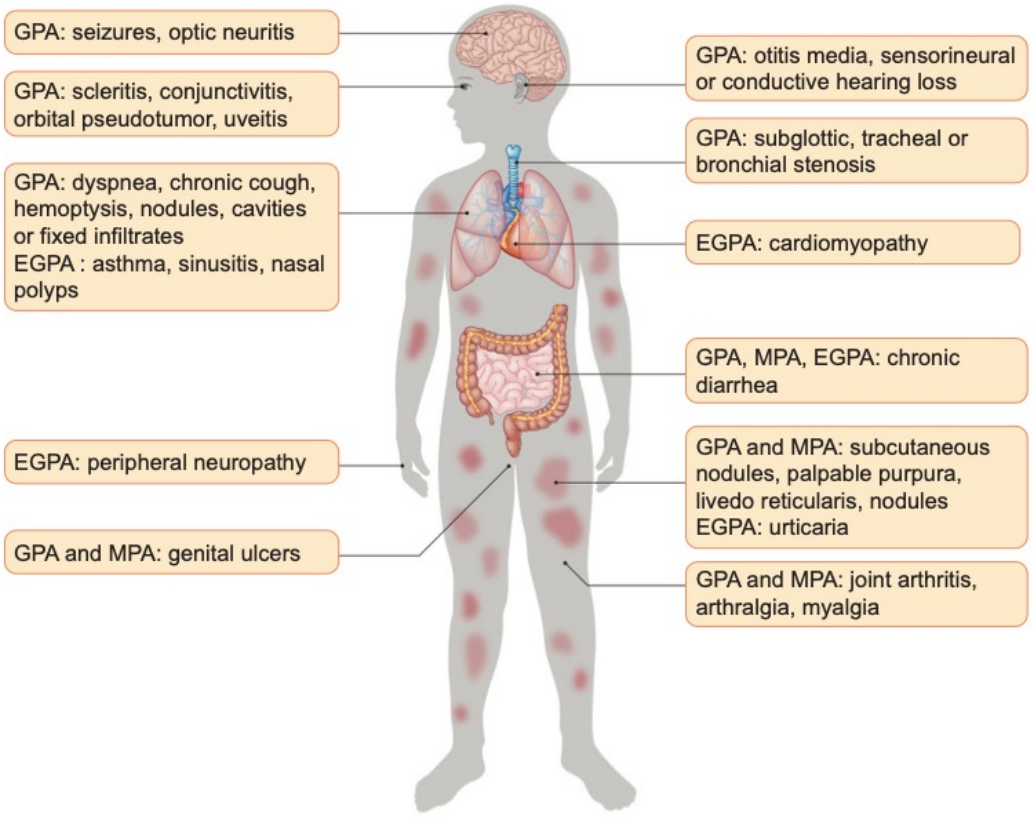
artpartner-images / Getty Images

96,500 – 160,000 km!

2.4 – 4 times the earth's circumference

~1/3 of the way to the moon

Small sized blood vessels are predominantly affected in childhood vasculitis, most frequently in the kidneys



Bernardi et al Nephrol Dial Transplant 2023 38:66-69

Story #1

See today. Predict tomorrow.

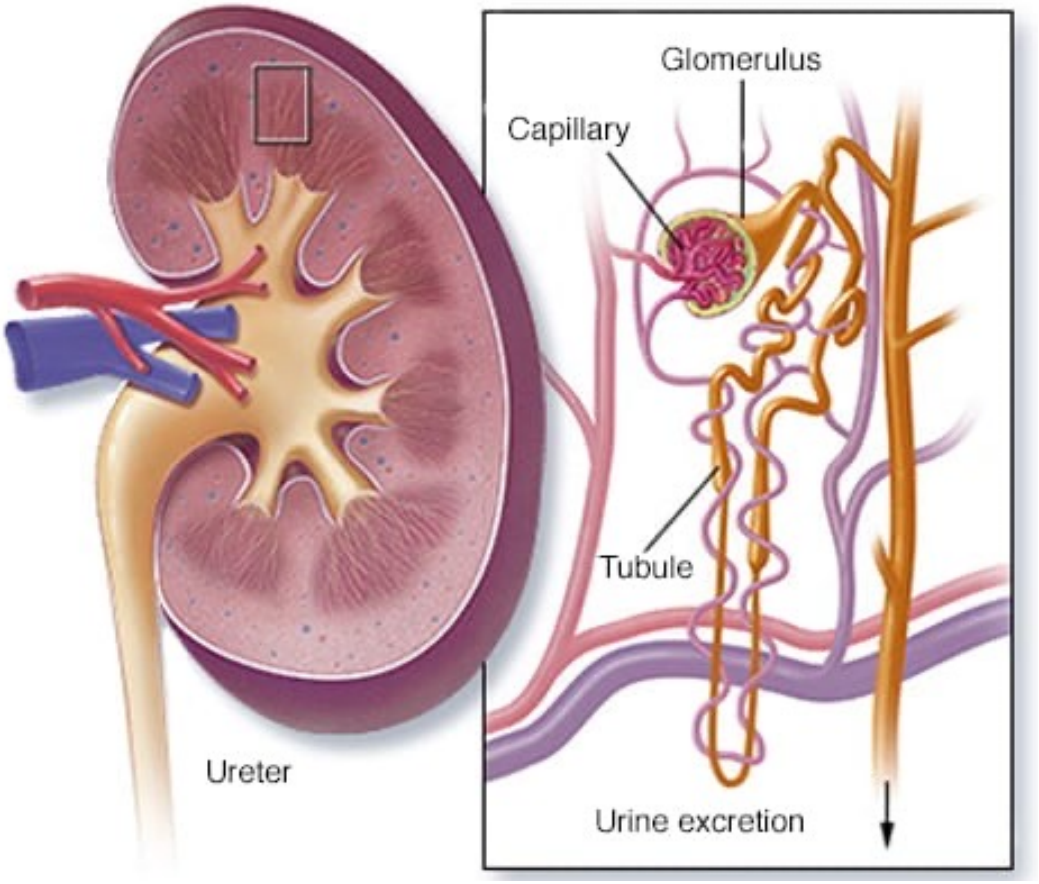


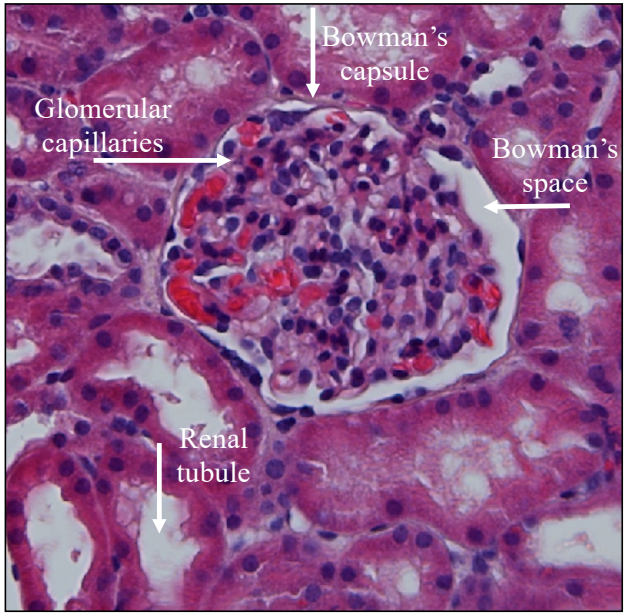
Image from Mayo Foundation for Medical Education and Research

Glomeruli are networks of tiny blood vessels and the main filtering unit of the kidneys.

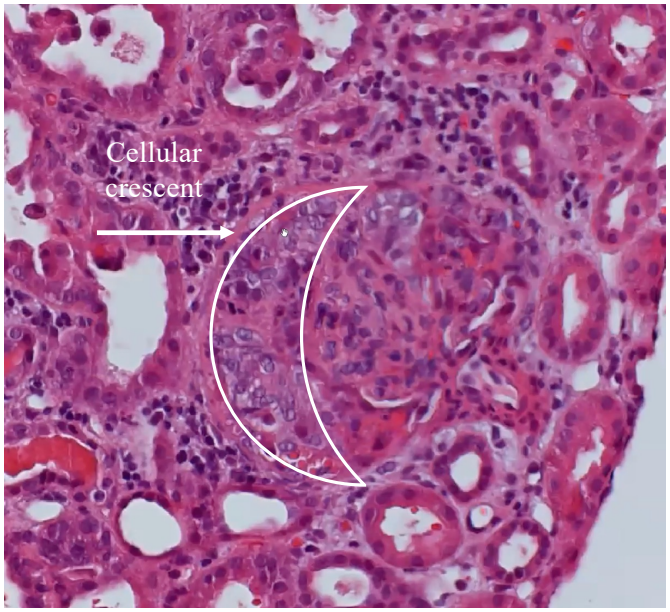
Glomerulonephritis is inflammation of the glomeruli.

Vasculitis-associated Glomerulonephritis

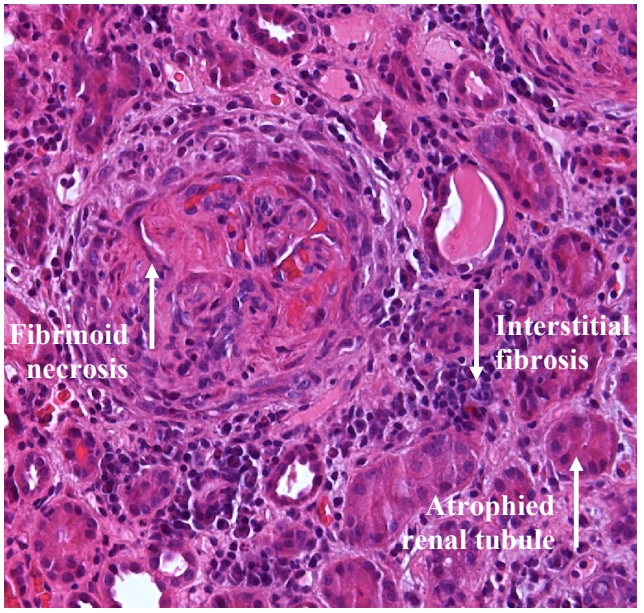
Healthy → Crescentic → Sclerotic



> 50%
good prognosis



> 50%
unpredictable
active (treatable) disease



> 50%
poor prognosis
permanent damage

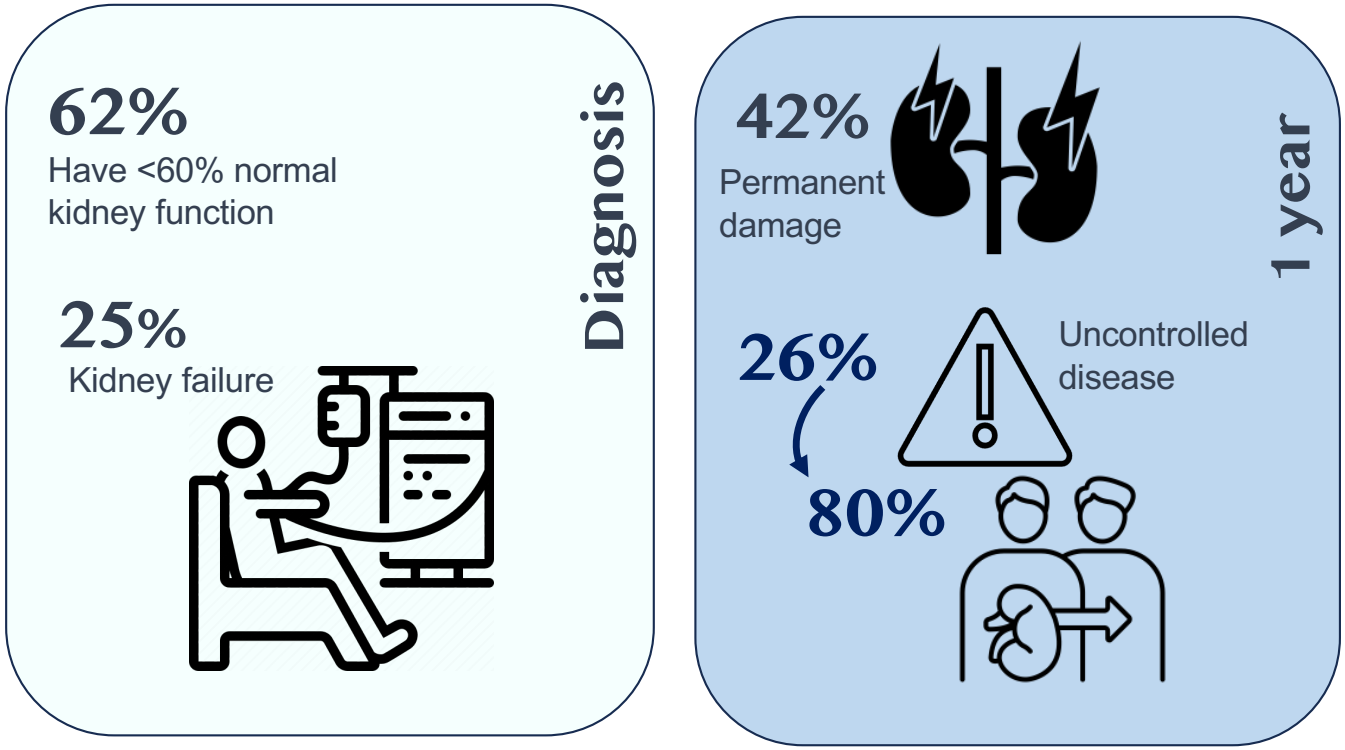
Biopsies from the Pediatric Vasculitis Initiative

Progression to end stage kidney disease can be rapid (weeks).

All patients get aggressive immune suppressive therapy at diagnosis

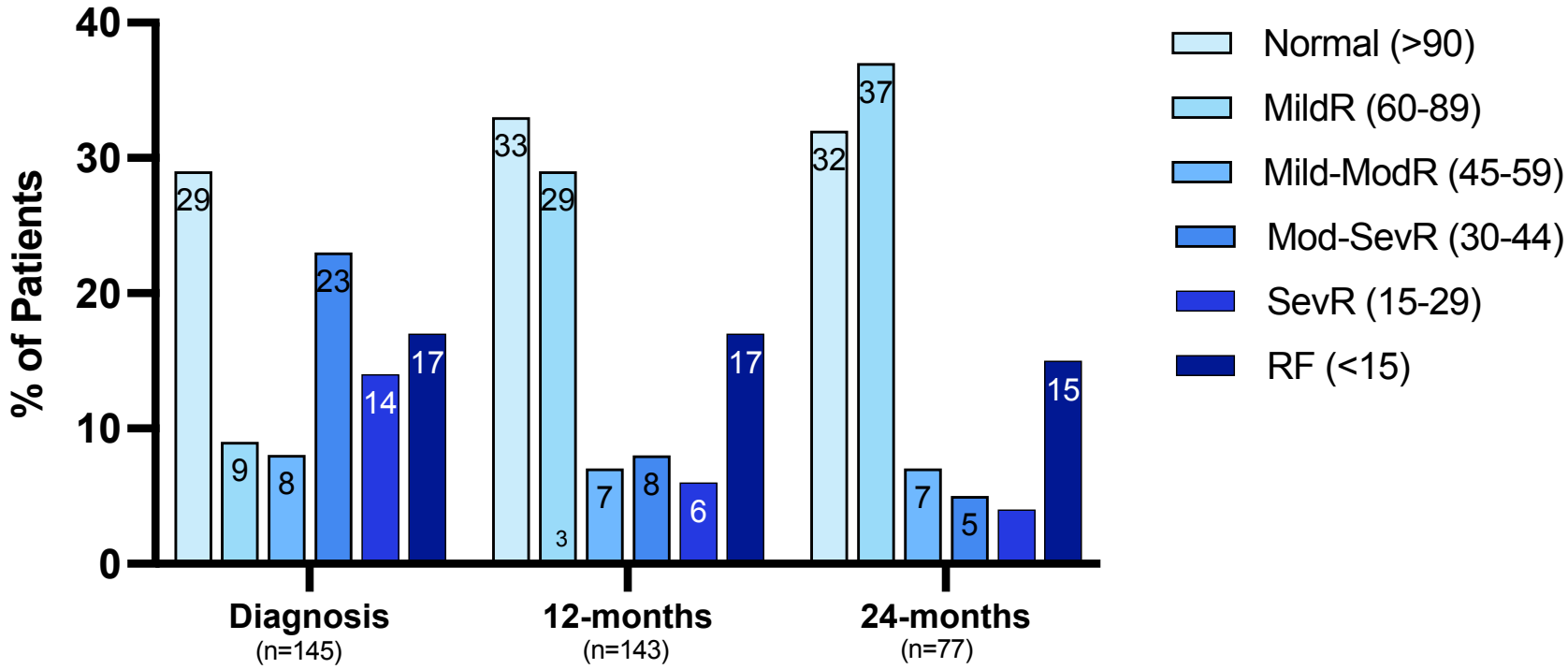
Goal is to recover or at least preserve existing kidney function

Vasculitis in the kidneys



Mann S et al RMD Open 2024 10:e004315 **n = 398 patients**
Toor K et al Arth Rheumatol 2025 77:606-614 **n = 145 patients**

Vasculitis in the kidneys



Andreas Kronbichler @AKronbichler · Jun 18, 2024

Another reason why ANCA-GN research is so important; children with MPO-ANCA vasculitis seem to have a better kidney function recovery than those with PR3-ANCA vasculitis. This is the other way around in adults!
Recent article in @bmj_rmdopen rmdopen.bmj.com/content/10/2/e...

Kidney Outcomes in Pediatric ANCA-Associated Vasculitis

https://doi.org/10.62716/kn.000292025

One year after diagnosis, most patients with pediatric-onset antineutrophil cytoplasmic antibody (ANCA)-associated vasculitis (AAV) have inactive kidney disease, but nearly half have evidence of kidney damage, reports a study in *Arthritis & Rheumatology*.

Using data from ARChiVe (A Registry of Childhood Vasculitis), the researchers identified 145 patients diagnosed with granulomatosis with polyangiitis, microscopic polyangiitis, or ANCA-positive pauci-immune glomerulonephritis. All were 18 years or younger at diagnosis, had kidney disease based on biopsy or dialysis dependence, and had clinical data at diagnosis and at a 12- or 24-month follow-up.

Rates of inactive kidney disease, defined as a pediatric vasculitis activity score of 0 or 1, were compared at 12 or 24 months. Improvements in kidney function and evidence of kidney damage were assessed at 24 months. The study also evaluated the prognostic implications of initial kidney function, based on estimated glomerular filtration rate (eGFR) and Kidney Disease: Improving Global Outcomes (KDIGO) stage.

Sixty-eight percent of patients were female; the median age at baseline was 13.8 years. The diagnosis was granulomatosis with polyangiitis in 78% of patients. Seventy-one percent of patients initially had less than normal kidney function, and 25% were on dialysis. At 12 months, 83% of patients with available data were classified as having inactive kidney disease. By 24 months, this figure increased to 98%.

However, 42% of patients had evidence of permanent kidney damage at 12 months. Findings included an eGFR of 15 to 60 mL/min/1.73 m^2 in 31% of patients, kidney failure with an eGFR less than 15 mL/min/1.73 m^2 in 21%, and hypertension over the 95th percentile or antihypertensive medication in 15%.

The baseline KDIGO category showed a linear association with follow-up outcomes. Odds ratios for a non-normal KDIGO category at 12 months were 4.77 for moderately reduced kidney function, 8.62 for severely reduced kidney function, and 26.3 for kidney failure at baseline. An eGFR of 38 mL/min/1.73 m^2 appeared to be the optimal cutoff for identifying patients at risk of moderate to severely reduced kidney function.

Pediatric-onset AAV is a rare, relapsing disease, with blood vessel damage potentially leading to downstream involvement of the kidneys and other organ systems. The new study adds to previous evidence of early kidney damage, even with aggressive treatment.

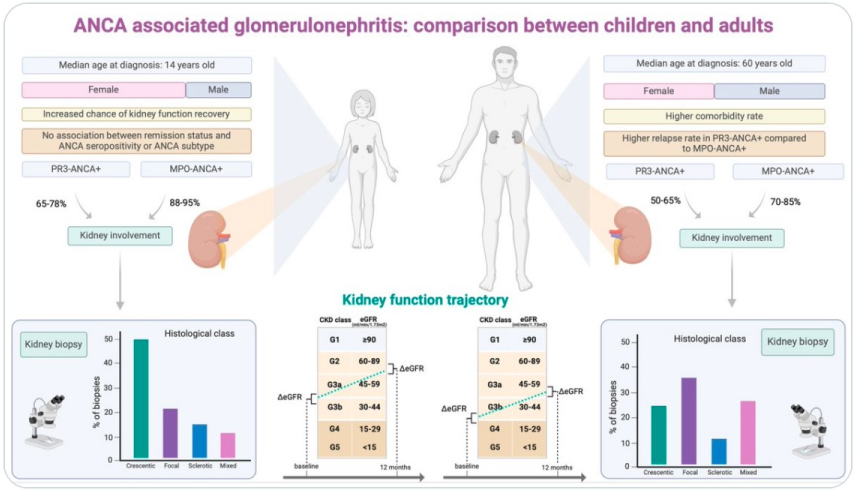
The extent of reduction in kidney function at baseline may provide a simple and practical tool for outcome prediction in pediatric AAV. "Providing improved, individualized insights into anticipated outcomes will ultimately allow patients and their families to be more involved in shaping and participating in their own care," the investigators conclude [Toor KK, et al.; PedVas Investigators Network. Evaluating renal disease in pediatric-onset antineutrophil cytoplasmic antibody-associated vasculitis: Disease course, outcomes, and predictors of outcome. *Arthritis Rheumatol*, published online December 3, 2024. doi: 10.1002/art.43071].

"If we can predict from an early stage how a child's disease will behave and how it will respond to treatment, then we can better tailor treatment."

Kimberly A. Morishita, MD, MHSc

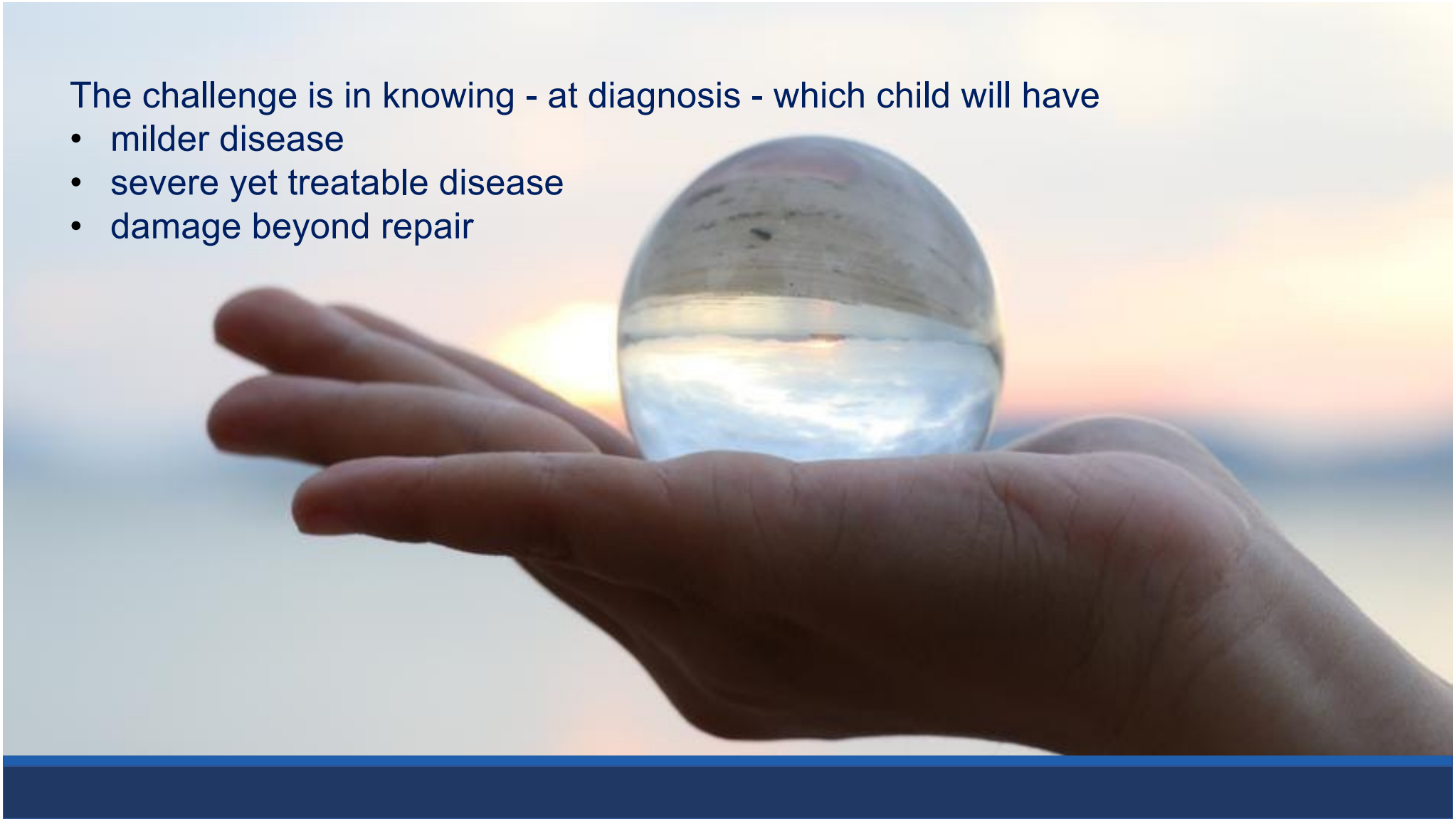
Andreas Kronbichler
@AKronbichler

Please find our Editorial discussing key differences between pediatric and adult ANCA-GN in @bmj_rmdopen!
rmdopen.bmj.com/content/10/3/e...
Together with @louise_oni and Dr. Barnini! Interesting aspects clearly highlighting huge differences ...



The challenge is in knowing - at diagnosis - which child will have

- milder disease
- severe yet treatable disease
- damage beyond repair



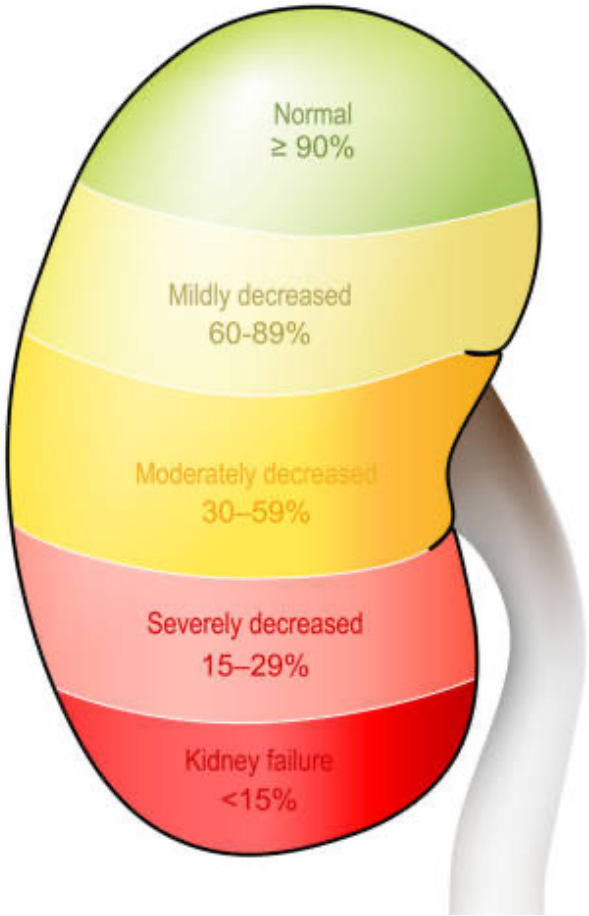
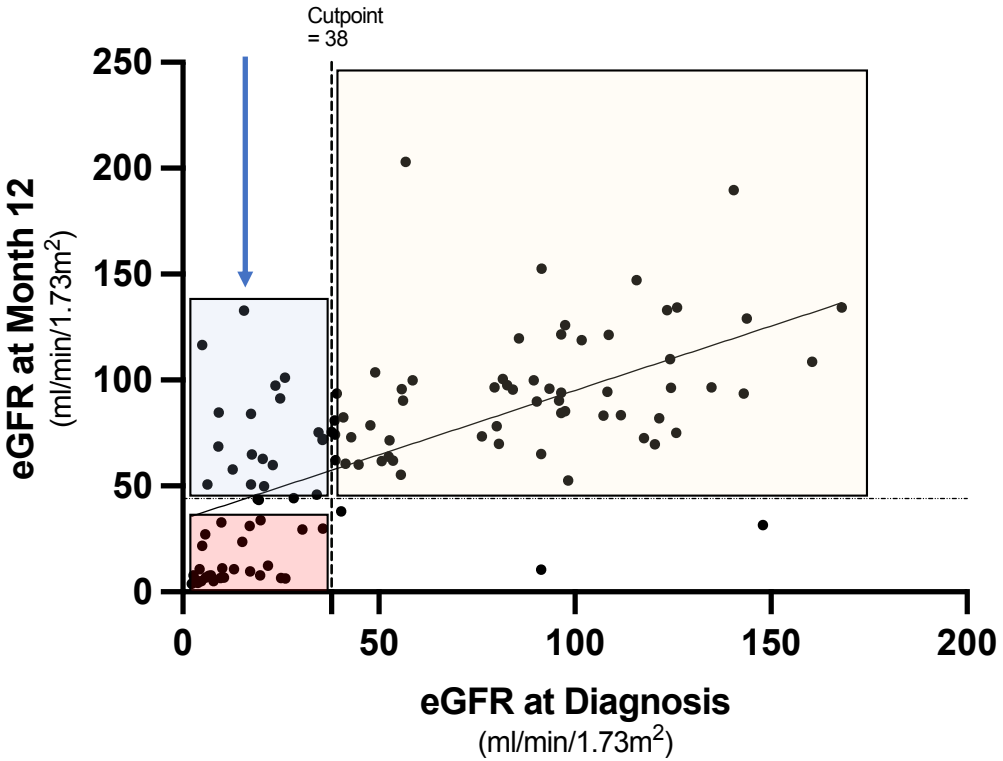


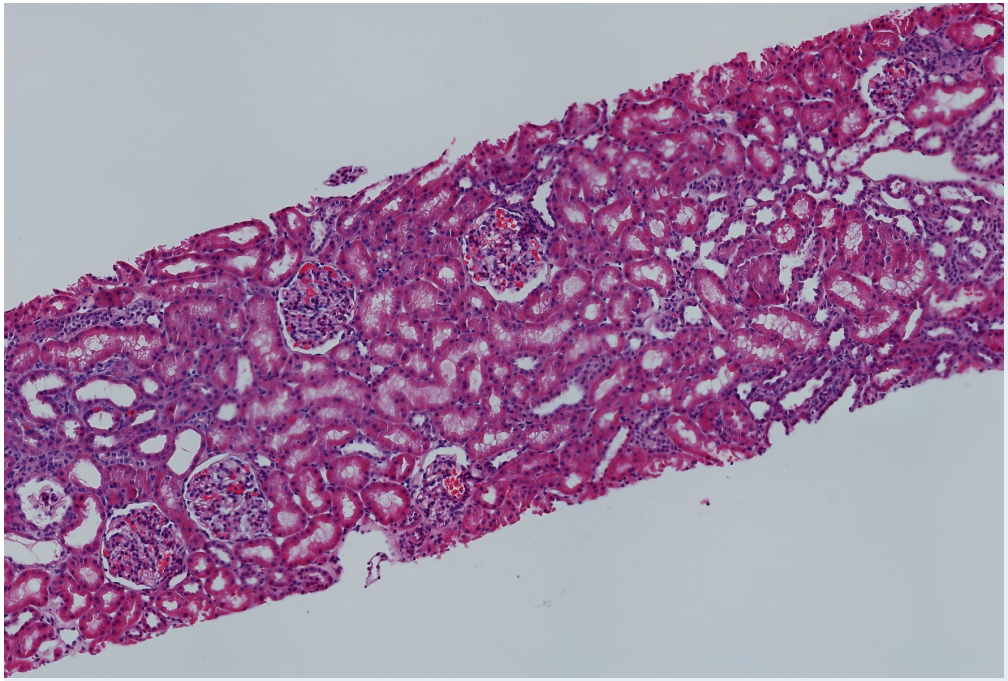
Image from iStock by Getty Images

*Estimated glomerular filtration rate (**eGFR**)* is the rate ($mL/min/1.73m^2$) that blood is filtered through the glomeruli.

It is a biomarker of kidney function.

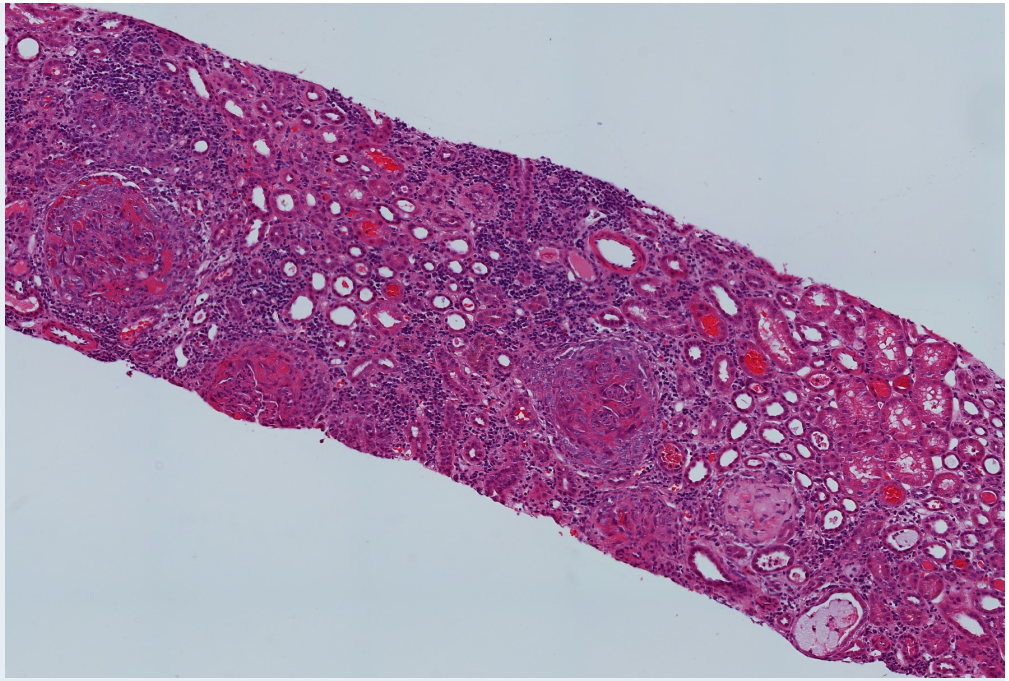
The need for Prognostic Biomarkers





@ diagnosis
normal function
healthy glomeruli

Did they maintain function because of the aggressive treatment?



@ diagnosis
kidney failure
crescentic glomeruli

Why did they not respond to treatment and need a transplant within 4 months?

Prognotic Biomarker Discovery