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Patient Voice

357 Responsive Designed Interventions Are Needed to Support Positive Outcomes of Children and Adolescents with CKD

Lori M. Hartwell

See related article on page 387.

Editorials

359 The Enigma of Blood Pressure Measurement in Children with CKD

Bonita Falkner

See related article on page 422.

361 Intensive Hemodialysis Fails to Reduce Plasma Levels of Uremic Solutes

Tammy L. Sirich and Timothy W. Meyer

See related article on page 436.

363 Rest Easy with Intravenous Iron for Dialysis Patients?: High Dose IV Iron Safety

Xiaojuan Li and Abhijit V. Kshirsagar

See related article on page 457.

Original Articles

Acid/Base and Electrolyte Disorders

366 Serum Sodium and Cognition in Older Community-Dwelling Men

Kristen L. Nowak, Kristine Yaffe, Eric S. Orwoll, Joachim H. Ix, Zhiying You, Elizabeth Barrett-Connor, Andrew R. Hoffman, and Michel Chonchol

Chronic Kidney Disease

375 Person-Centered Integrated Care for Chronic Kidney Disease: A Systematic Review and Meta-Analysis of Randomized Controlled Trials

Pim P. Valentijn, Fernando Abdalla Pereira, Marinella Ruospo, Suetonia C. Palmer, Jörgen Hegbrant, Christina W. Sterner, Hubertus J.M. Vrijhoef, Dirk Ruwaard, and Giovanni F.M. Strippoli

Clinical Nephrology

387 Neurocognitive and Educational Outcomes in Children and Adolescents with CKD: A Systematic Review and Meta-Analysis

Kerry Chen, Madeleine Didsbury, Anita van Zwieten, Martin Howell, Siah Kim, Allison Tong, Kirsten Howard, Natasha Nassar, Belinda Barton, Suncica Lah, Jennifer Lorenzo, Giovanni Strippoli, Suetonia Palmer, Armando Teixeira-Pinto, Fiona Mackie, Steven McTaggart, Amanda Walker, Tonya Kara, Jonathan C. Craig, and Germaine Wong

See related Patient Voice on page 357.

Diabetes and the Kidney

398 Kidney Biomarkers and Decline in eGFR in Patients with Type 2 Diabetes

Katherine G. Garlo, William B. White, George L. Bakris, Faiez Zannad, Craig A. Wilson, Stuart Kupfer, Muthiah Vaduganathan, David A. Morrow, Christopher P. Cannon, and David M. Charytan

Glomerular and Tubulointerstitial Diseases

- 406 Mycophenolate Mofetil in Combination with Steroids for Treatment of C3 Glomerulopathy: A Case Series**
Rupali S. Avasare, Pietro A. Canetta, Andrew S. Bomback, Maddalena Marasa, Yasar Caliskan, Yasemin Ozluk, Yifu Li, Ali G. Gharavi, and Gerald B. Appel
- 414 An Outcomes-Based Definition of Proteinuria Remission in Focal Segmental Glomerulosclerosis**
Jonathan P. Troost, Howard Trachtman, Patrick H. Nachman, Matthias Kretzler, Cathie Spino, Radko Komers, Sarah Tuller, Kalyani Perumal, Susan F. Massengill, Elaine S. Kamil, Gia Oh, David T. Selewski, Patrick Gipson, and Debbie S. Gipson

Hypertension

- 422 Twenty-Four-Hour Ambulatory Blood Pressure versus Clinic Blood Pressure Measurements and Risk of Adverse Outcomes in Children with CKD**
Elaine Ku, Charles E. McCulloch, Bradley A. Warady, Susan L. Furth, Barbara A. Grimes, and Mark M. Mitsnefes
See related editorial on page 359.

Maintenance Dialysis

- 429 Effectiveness and Cost of Weekly Recombinant Tissue Plasminogen Activator Hemodialysis Catheter Locking Solution**
Brenda R. Hemmelgarn, Braden J. Manns, Steven D. Soroka, Adeera Levin, Jennifer MacRae, Karthik Tennankore, Jo-Anne S. Wilson, Robert G. Weaver, Pietro Ravani, Robert R. Quinn, Marcello Tonelli, Mercedeh Kiaii, Paula Mossop, and Nairne Scott-Douglas
- 436 Extended Duration Nocturnal Hemodialysis and Changes in Plasma Metabolite Profiles**
Sahir Kalim, Ron Wald, Andrew T. Yan, Marc B. Goldstein, Mercedeh Kiaii, Dihua Xu, Anders H. Berg, Clary Clish, Ravi Thadhani, Eugene P. Rhee, and Jeffrey Perl
See related editorial on page 361.
- 445 Weekly Standard Kt/V_{urea} and Clinical Outcomes in Home and In-Center Hemodialysis**
Matthew B. Rivara, Vanessa Ravel, Elani Streja, Yoshitsugu Obi, Melissa Soohoo, Alfred K. Cheung, Jonathan Himmelfarb, Kamyar Kalantar-Zadeh, and Rajnish Mehrotra
- 457 Safety of Intravenous Iron in Dialysis: A Systematic Review and Meta-Analysis**
Ingrid Hougen, David Collister, Mathieu Bourrier, Thomas Ferguson, Laura Hochheim, Paul Komenda, Claudio Rigatto, and Navdeep Tangri
See related editorial on page 363.

Perspectives

- 468 Striving to Achieve an Integrated Home Dialysis System: A Report from the Ontario Renal Network Home Dialysis Attrition Task Force**
Brendan B. McCormick and Christopher T. Chan for the ORN Home Dialysis Research Group
- 471 A View of the Bundle from a Home Dialysis Perspective: Present at the Creation**
Thomas A. Golper
See related article on page 474.
- 474 The Dialysis Facility Compare Five-Star Rating System at 2 Years**
Alyssa Pozniak and Jeffrey Pearson
See related article on page 471.
- 477 Need to Reclassify Etiologies of ESRD on the CMS 2728 Medical Evidence Report**
Bryan M. Tucker and Barry I. Freedman
- 480 The Affordable Care Act, Medicaid Expansion, and Disparities in Kidney Disease**
Amal N. Trivedi and Benjamin D. Sommers

Kidney Case Conference: How I Treat

- 483 Crafting the Prescription for Patients Starting Peritoneal Dialysis**
Isaac Teitelbaum

486 **Diagnosis and Treatment of Intradialytic Hypotension in Maintenance Hemodialysis Patients**

Christopher W. McIntyre and Fabio R. Salerno

Moving Points in Nephrology

490 **Clinical Trial End Points for Hemodialysis Vascular Access: Background, Rationale, and Definitions**

Surendra Shenoy, Michael Allon, Gerald Beathard, Deborah Brouwer-Maier, Laura M. Dember, Mark Glickman, Celeste Lee, Terry Litchfield, Charmaine Lok, Thomas Huber, Prabir Roy-Chaudhury, Jack Work, Melissa West, and Haimanot Wasse

See related articles on pages 495, 501 and 513.

495 **Recommended Clinical Trial End Points for Dialysis Catheters**

Michael Allon, Deborah J. Brouwer-Maier, Kenneth Abreo, Kevin M. Baskin, Kay Bregel, Deepa H. Chand, Andrea M. Easom, Leonard Mermel, Michele H. Mokrzycki, Priti R. Patel, Prabir Roy-Chaudhury, Surendra Shenoy, Rudolph P. Valentini, and Haimanot Wasse

See related articles on pages 490, 501 and 513.

501 **Definitions and End Points for Interventional Studies for Arteriovenous Dialysis Access**

Gerald A. Beathard, Charmaine E. Lok, Marc H. Glickman, Ahmed A. Al-Jaishi, Donna Bednarski, David L. Cull, Jeffery H. Lawson, Timmy C. Lee, Vandana D. Niyyar, Donna Syracuse, Scott O. Trerotola, Prabir Roy-Chaudhury, Surendra Shenoy, Margo Underwood, Haimanot Wasse, Karen Woo, Theodore H. Yuo, and Thomas S. Huber

See related articles on pages 490, 495 and 513.

513 **FDA Regulatory Perspectives for Studies on Hemodialysis Vascular Access**

Frank P. Hurst, Robert E. Lee, Aliza M. Thompson, Brian D. Pullin, and Douglas M. Silverstein

See related articles on pages 490, 495 and 501.

On the Cover

What's the diagnosis?

A 53-year-old man presented with progressive lower extremity edema and acute kidney injury with serum creatinine 9.6 mg/dl (baseline 2.5 mg/dl). Urinalysis/urine sediment was strongly positive for proteinuria (3+) and mild hematuria without dysmorphic RBCs or RBCs casts. Twenty-four-hour urine protein was 7.1 g. Kidney ultrasound was normal. AntiPLA2R antibody assay was strongly positive with a titer of 1500 relative units/ml. Complement levels were normal. All other serologies were negative with no monoclonal gammopathy and negative malignancy workup. Kidney biopsy demonstrated evidence of membranous nephropathy along with crescents. The patient was diagnosed with primary crescentic membranous nephropathy. He was started on lisinopril along with prednisone monotherapy. Patient declined using any other immunosuppression agents. The patient's serum creatinine returned to baseline and protein/creatinine ratio improved to 2.4 g/24 hr within four months.

Light microscopy showed mesangial matrix expansion, thickened of the capillary walls, and active cellular crescents (Figures 1 and 2). Electron microscopy demonstrated diffuse effacement of the podocyte foot processes involving 80—90% of the capillary surfaces with subepithelial deposits (Figure 2).

Primary crescentic membranous nephropathy with negative serology is a rare disease and was reported in only a handful of prior case report/series.¹ Patients usually present with nephrotic range proteinuria, hematuria and acute decline in kidney function. Kidney biopsy confirms the diagnosis. Currently there are no guidelines regarding the ideal treatment in these cases. Unlike typical membranous nephropathy, crescentic membranous nephropathy usually progresses long-term, even if patients experience initial positive response to therapy.

(Images and text provided by Mohamad Hanouneh, MD, Steven Menez, and Duwuru Geetha, MBBS, MD, Johns Hopkins University, Department of Medicine, Baltimore, Maryland)

Reference

1. Rodriguez EF, Nasr SH, Larsen CP, Sethi S, Fidler ME, Cornell LD: Membranous nephropathy with crescents: a series of 19 cases. *Am J Kidney Dis* 64: 66–73, 2014