

CJASN

Clinical Journal of the American Society of Nephrology

January 2020 • Vol. 15 • No. 1

Patient Voice

1 Disparities in Health Outcomes with Dialysis in the United States Vary by Race

Daronta L. Briggs

See related article on page 101.

Editorials

2 Renin-Angiotensin System Blockade after Acute Kidney Injury: The Plot Thickens

Steven Menez and Chirag R. Parikh

See related article on page 26.

5 Lipids and Cardiovascular Risk with CKD

Farsad Afshinnia and Subramaniam Pennathur

See related article on page 47.

8 Keep Children with CKD Safe from Inappropriate Prescribing

Stuart L. Goldstein

See related article on page 61.

10 "It's In Your Genes": Exome Sequencing Enables Precision Diagnostics in Proteinuric Kidney Diseases

Franz Schaefer

See related article on page 89.

13 Medical Therapies to Reduce Delayed Graft Function and Improve Long-Term Graft Survival: Are We Making Progress?

Mary Ann Lim and Roy D. Bloom

See related article on page 109.

Original Articles

Acid/Base and Electrolyte Disorders

16 Tubular Acidification Defect in Adults with Sickle Cell Disease

Maud Cazenave, Vincent Audard, Jean-Philippe Bertocchio, Anoosha Habibi, Stéphanie Baron, Caroline Prot-Bertoye, Jugurtha Berkenou, Gérard Maruani, Thomas Stehlé, Nicolas Cornière, Hamza Ayari, Gérard Friedlander, Frédéric Galacteros, Pascal Houillier, Pablo Bartolucci, and Marie Courbebaisse

Acute Kidney Injury and ICU Nephrology

26 Renin-Angiotensin System Blockade after Acute Kidney Injury (AKI) and Risk of Recurrent AKI

Chi-yuan Hsu, Kathleen D. Liu, Jingrong Yang, David V. Glidden, Thida C. Tan, Leonid Pravoverov, Sijie Zheng, and Alan S. Go

See related editorial on page 2.

35 Intraoperative Arterial Pressure Variability and Postoperative Acute Kidney Injury

Sehoon Park, Hyung-Chul Lee, Chul-Woo Jung, Yunhee Choi, Hyung Jin Yoon, Sejoong Kim, Ho Jun Chin, Myoungsuk Kim, Yong Chul Kim, Dong Ki Kim, Kwon Wook Joo, Yon Su Kim, and Hajeong Lee

Chronic Kidney Disease

47 Apolipoprotein B, Triglyceride-Rich Lipoproteins, and Risk of Cardiovascular Events in Persons with CKD

Julio Alejandro Lamprea-Montealegre, Natalie Staplin, William G. Herrington, Richard Haynes, Jonathan Emberson, Colin Baigent, and Ian H. de Boer, on behalf of the SHARP Collaborative Group
See related editorial on page 5.

61 Primary Care Prescriptions of Potentially Nephrotoxic Medications in Children with CKD

Claire E. Lefebvre, Kristian B. Filion, Pauline Reynier, Robert W. Platt, and Michael Zappitelli
See related editorial on page 8.

69 Epidemiology of Autosomal Dominant Polycystic Kidney Disease in Olmsted County

Tatsuya Suwabe, Shehbaz Shukoor, Alanna M. Chamberlain, Jill M. Killian, Bernard F. King, Marie Edwards, Sarah R. Senum, Charles D. Madsen, Fouad T. Chebib, Marie C. Hogan, Emilie Cornec-Le Gall, Peter C. Harris, and Vicente E. Torres

Cystic Kidney Disease

80 Alterations of Proximal Tubular Secretion in Autosomal Dominant Polycystic Kidney Disease

Ke Wang, Leila R. Zelnick, Yan Chen, Andrew N. Hoofnagle, Terry Watnick, Stephen Seliger, and Bryan Kestenbaum

Genetics

89 Reverse Phenotyping after Whole-Exome Sequencing in Steroid-Resistant Nephrotic Syndrome

Samuela Landini, Benedetta Mazzinghi, Francesca Becherucci, Marco Allinovi, Aldesia Provenzano, Viviana Palazzo, Fiammetta Ravaglia, Rosangela Artuso, Emanuele Bosi, Stefano Stagi, Giulia Sansavini, Francesco Guzzi, Luigi Cirillo, Augusto Vaglio, Luisa Murer, Licia Peruzzi, Andrea Pasini, Marco Materassi, Rosa Maria Roperto, Hans-Joachim Anders, Mario Rotondi, Sabrina Rita Giglio, and Paola Romagnani
See related editorial on page 10.

Maintenance Dialysis

101 Racial and Ethnic Variations in Mortality Rates for Patients Undergoing Maintenance Dialysis Treated in US Territories Compared with the US 50 States

Guofen Yan, Jenny I. Shen, Rubette Harford, Wei Yu, Robert Nee, Mary Jo Clark, Jose Flaque, Jose Colon, Francisco Torre, Ylene Rodriguez, Jane Georges, Lawrence Agodoa, and Keith C. Norris
See related Patient Voice on page 1.

Transplantation

109 Three-Year Outcomes of a Randomized, Double-Blind, Placebo-Controlled Study Assessing Safety and Efficacy of C1 Esterase Inhibitor for Prevention of Delayed Graft Function in Deceased Donor Kidney Transplant Recipients

Edmund Huang, Ashley Vo, Jua Choi, Noriko Ammerman, Kathlyn Lim, Supreet Sethi, Irene Kim, Sanjeev Kumar, Riad Najjar, Alice Peng, and Stanley C. Jordan
See related editorial on page 13.

Research Letter

117 Exclusion of Persons with Kidney Disease in Trials of Peripheral Artery Disease: A Systematic Review of Randomized Trials

Yumeng Wen, O. Alison Potok, Xin Wei, Ishan Paranjpe, Lili Chan, Rakesh Malhotra, Steven Coca, Jagat Narula, Jeffrey W. Olin, Girish N. Nadkarni, and Pranav S. Garimella

Kidney Case Conference: How I Treat

120 Pregnancy in a Kidney Transplant Patient

Song C. Ong and Vineeta Kumar

Perspectives

123 Prophylactic Anticoagulation in Adult Patients with Nephrotic Syndrome

Judit Gordon-Cappitelli and Michael J. Choi

126 Genetic Testing for *APOL1* Genetic Variants in Clinical Practice: Finally Starting to Arrive

Jeffrey B. Kopp and Cheryl A. Winkler

129 Preserving Kidney Function Instead of Replacing It

Alan S. Kliger and Frank C. Brosius, on behalf of the Diabetic Kidney Disease Task Force of the American Society of Nephrology

Reviews

132 Prevention of Bloodstream Infections in Patients Undergoing Hemodialysis

Molly Fisher, Ladan Golestaneh, Michael Allon, Kenneth Abreo, and Michele H. Mokrzycki

152 Ambulatory Hemodialysis-Technology Landscape and Potential for Patient-Centered Treatment

Nina Hojs, William H. Fissell, and Shuvo Roy

On the Cover

What's the diagnosis?

In 2014, a 37-year-old white man previously healthy presented with an increasing abdominal girth over two weeks and blood pressure of 160/85 mmHg. Physical exam was significant for abdominal distention with dullness to percussion. Ultrasound confirmed ascites and paracentesis was consistent with exudative fluid. Computed tomography (CT) scan revealed extensive peritoneal and omental thickening with diffusely enlarged kidneys (Figure 1). Ultrasound-guided omental, peritoneal, and renal biopsies were diagnostic of a sclerosing fibro-inflammatory proliferation with foamy histiocytic infiltrate. Histopathology was reviewed and verified by three independent laboratories. The cellular infiltrate was considered nonmalignant, and no specific treatment was initiated, and the lesions abated spontaneously. During the clinic follow up, his hypertension workup included serum aldosterone and renin activity which were elevated at 30.6 ng/dl and 14.2 ng/ml/hr respectively, when his blood pressure was 173/87 mmHg; suggestive of renovascular-mediated hypertension. His repeat CT scan showed interval development of subcapsular densities surrounding both kidneys. A diagnosis of secondary hypertension due to Page kidney process was made, and spironolactone was initiated, which controlled his blood pressure to 133-144/76-85 mmHg (Table 1). Page kidney is analogous to the renal wrap hypertension model that is also renin-dependent [1,2]. He sustained recurrent acute kidney injuries due to partial obstruction, primarily due to the encapsulation of the renal pelves and ureters that required ureteral stenting. In these instances, spironolactone was discontinued due to the obstruction-associated hyperkalemia; his blood pressure needed multiple high doses of alternative anti-hypertension medications to achieve comparable blood pressure control. In June 2015, the infiltrate recurred, and the histology was deemed to have developed into more aggressive pre-malignant form. He was treated with prednisone with stabilization. He remained on spironolactone until late 2017, but due to recurrent episodes of acute kidney injury with hyperkalemia and progressive loss of function (eGFR in 2019, 33 ml/min per 1.73m²), his blood pressure has been treated with less-than-optimal success on four medications plus a diuretic. (Cover Image and text provided by Nashat Imran, Internal Medicine Department, Nephrology Division, Assistant Professor, Wayne State University; Nadine Abdallah, Internal Medicine Department, Medical Resident, Wayne State University; and Noreen F. Rossi; Internal Medicine Department, Nephrology Division, Professor, Wayne State University.)

References

1. Denton KM, Anderson WP: Role of angiotensin II in renal wrap hypertension. *Hypertension* 7(6 Pt 1): 893-898, 1985
2. Dopson SJ, Jayakumar S, Velez JC: Page kidney as a rare cause of hypertension: case report and review of the literature. *Am J Kidney Dis* 54(2): 334-339, 2009

Table 1. Renin and Aldosterone Levels and Treatment Effect on Outpatient Blood Pressure over Time

	Systolic	Diastolic	Renin ng/ml/hr	Aldosterone ng/dl	Spironolactone (on/off)
May 2014	173	87	14.2	30.6	Time of diagnosis (off)
July 2014	144	85			on
November 2014	133	76			on
March 2015	140	80			on
June 2015	155	91			off
December 2015	154	90	6.1	38.3	off
December 2016	151	85			on
August 2017	145	85			on
July 2018	137	71			on
January 2019	173	97			off