Patient Voice

1073 Long-Term Hemodialysis during the COVID-19 Pandemic
Nieltje Gedney
See related article on page 1139.

Editorials

1075 Adverse Drug Effects in Patients with CKD: Primum Non Nocere
Mark A. Perazella and Thomas D. Nolin
See related article on page 1090.

1078 Keeping Up with the Times: Prognostic Tools in ANCA-Associated Glomerulonephritis
Silke R. Brix and Duvuru Geetha
See related article on page 1103.

1081 Intensive Blood Pressure Lowering Should Be the Goal for Most Individuals at High Risk of Cardiovascular Disease Irrespective of Albuminuria
Simon B. Ascher and Joachim H. Ix
See related article on page 1121.

1084 Drug Selection for Treating Hypertension in Dialysis Patients: More to Consider than BP-Lowering Potency
Tariq Shafi and Dana C. Miskulin
See related article on page 1129.

1087 COVID-19 in Patients with Kidney Disease
Maria Ajaimy and Michal L. Melamed
See related articles on pages 1139 and 1174.

Original Articles

Chronic Kidney Disease

1090 Adverse Drug Reactions in Patients with CKD
Solène M. Laville, Valérie Gras-Champel, Julien Moragny, Marie Metzger, Christian Jacquelinet, Christian Combe, Denis Fouque, Maurice Laville, Luc Frimat, Bruce M. Robinson, Bénédicte Stengel, Ziad A. Massy, and Sophie Liabeuf, on behalf of the Chronic Kidney Disease-Renal Epidemiology and Information Network (CKD-REIN) Study Group
See related editorial on page 1075.

Glomerular and Tubulointerstitial Diseases

1103 Developments in the Histopathological Classification of ANCA-Associated Glomerulonephritis
Emma E. van Daalen, Maria A.C. Wester Trejo, Arda Göçeroğlu, Franco Ferrario, Kensuke Joh, Laure-Hélène Noël, Yayoi Ogawa, Suzanne Wilhelmus, Miriam J. Ball, Eva Honsova, Zdenka Hruskova, Renate Kain, Tomoyoshi Kimura, Marek Kollar, Andreas Kronbichler, Kristine Lindhard, Xavier Puéchal, Steven Salvatore, Wladimir Szpirt, Hideki Takizawa, Vladimir Tesar, Annelies E. Berden, Olaf M. Dekkers, E. Christiaan Hagen, Jan Oosting, Chinar Rahmatulla, Ron Wolterbeek, Willem Jan Bos, Jan A. Bruijn, and Ingeborg M. Bajema
See related editorial on page 1078.
Glomerular and Tubulointerstitial Diseases (Continued)

1112 External Validation of the International IgA Nephropathy Prediction Tool
Junjun Zhang, Bo Huang, Zhangsuo Liu, Xutong Wang, Minhua Xie, Ruxue Guo, Yongli Wang, Dan Yu, Panfeng Wang, Yuzhe Zhu, and Jingjing Ren

Hypertension

1121 Effects of Intensive Blood Pressure Control in Patients with and without Albuminuria: Post Hoc Analyses from SPRINT
Alex R. Chang, Holly Kramer, Guo Wei, Robert Boucher, Morgan E. Grams, Dan Berlowitz, Udayan Bhatt, Debbie L. Cohen, Paul Drawz, Henry Punzi, Barry I. Freedman, William Haley, Amret Hawfield, Edward Horwitz, Christopher McLouth, Don Morisky, Vasilios Papademetriou, Michael V. Rocco, Barry Wall, Daniel E. Weiner, Athena Zias, and Srinivasan Beddu, for the SPRINT Research Group
See related editorial on page 1081.

Maintenance Dialysis

1129 Comparative Efficacy and Safety of BP-Lowering Pharmacotherapy in Patients Undergoing Maintenance Dialysis: A Network Meta-Analysis of Randomized, Controlled Trials
See related editorial on page 1084.

1139 Clinical Features of Maintenance Hemodialysis Patients with 2019 Novel Coronavirus-Infected Pneumonia in Wuhan, China
Jun Wu, Jushuang Li, Geli Zhu, Yanxia Zhang, Zhimin Bi, Yean Yu, Bo Huang, Shouzhi Fu, Yiqing Tan, Jianbin Sun, and Xiangyou Li
See related Patient Voice and editorial on pages 1073 and 1087, respectively.

1146 Apixaban versus No Anticoagulation in Patients Undergoing Long-Term Dialysis with Incident Atrial Fibrillation
Thomas A. Mavrakanas, Katherine Garlo, and David M. Charytan

Nephrolithiasis

1155 Efficacy and Safety of Daprodustat Compared with Darbepoetin Alfa in Japanese Hemodialysis Patients with Anemia: A Randomized, Double-Blind, Phase 3 Trial
Tadao Akizawa, Masaomi Nangaku, Taeko Yonekawa, Nobuhiko Okuda, Shinya Kawamatsu, Tomohiro Onoue, Yukihiro Endo, Katsutoshi Hara, and Alexander R. Cobitz

Transplantation

1174 Early Outcomes of Outpatient Management of Kidney Transplant Recipients with Coronavirus Disease 2019
See related editorial on page 1087.

Research Letter

1179 Association of Race and Risk of Graft Loss among Kidney Transplant Recipients in the US Military Health System
Crystal J. Forman, Christina M. Yuan, Rahul M. Jindal, Lawrence Y. Agodoa, Kevin C. Abbott, and Robert Nee
Genomics of Kidney Disease

1181 At the Research-Clinical Interface: Returning Individual Genetic Results to Research Participants
Kathleen M. West, Erika Blacksher, Kerri L. Cavanaugh, Stephanie M. Fullerton, Ebele M. Umeukeje, Bessie A. Young, and Wylie Burke

Kidney Case Conference: Nephrology Quiz and Questionnaire

1190 Use of Checkpoint Inhibitors in a Kidney Transplant Recipient with Metastatic Cancer
Song Ching Ong and Roslyn B. Mannon

Perspectives

1193 I Love Nephrology, but Should I Be a Nephrologist? An Early Career Trainee Perspective
Tanima Arora

1195 Ensuring Gender-Affirming Care in Nephrology: Improving Care for Transgender and Gender-Expansive Individuals
Dinushika Mohottige and Mitchell R. Lunn

1198 Careers in Critical Care Nephrology: Opportunities and Challenges
Sarah F. Sanghavi and Kirk N. Campbell

1201 Precision in GFR Reporting: Let’s Stop Playing the Race Card
Vanessa Grubbs
See related article on page 1203.

Features

1203 Kidney Disease, Race, and GFR Estimation
Andrew S. Levey, Silvia M. Titan, Neil R. Powe, Josef Coresh, and Lesley A. Inker
See related Perspective on page 1201.

1213 Presidential Address Kidney Week 2019: Winning the War on Kidney Diseases: The Time Is Now
Mark E. Rosenberg

On the Cover

What is the diagnosis?
A 32-year-old male presented for initial evaluation of CKD with a history significant for Fabry disease. Past medical history was notable for irritable bowel syndrome, painful neuropathy of his extremities, and anhidrosis. On examination, blood pressure was 110/74 mmHg; there was no flank tenderness. Serum creatinine was 0.64 mg/dl. Urine dipstick was positive for protein of 100mg/dl but otherwise bland.

Image Description:
A screening renal ultrasound showed multiple anechoic lesions in the renal sinus with no connection to the ureter, which was initially characterized as moderate bilateral hydronephrosis (Left). Magnetic resonance imaging (MRI) with gadolinium was performed to further evaluate the presence of obstruction. This revealed multiple bilateral T2 hyperintense parapelvic cysts without communication with the collecting system (Center). These were non-enhancing cystic lesions along the renal sinuses without evidence of contrast filling on T1 delayed postcontrast imaging (Right). There was no obstructing calculus or hydronephrosis, and the corresponding findings on the ultrasound were attributed to the bilateral renal sinus cysts discovered on MRI.

Teaching Points:
Parapelvic cysts are common in Fabry disease, with an estimated prevalence of 29%–43%, significantly higher than the approximately 1% estimated prevalence in the general population; but they are not specific to this condition. They are often misdiagnosed as hydronephrosis due to the hypoechoic character by ultrasonography and the location in the renal sinus. When asymptomatic hydronephrosis is noted by ultrasonography without identifiable obstructive lesions in patients with Fabry disease, the possibility of parapelvic cysts should be considered.

Disclaimer:
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(Images and text provided by Richard Plasse, DO, Meghla Joshi, DO, Robert Nee, MD, and Maura Watson, DO, MPH, Nephrology Service, Walter Reed National Military Medical Center, Bethesda, MD, and Department of Medicine, Uniformed Services University, Bethesda, MD; and Nathan Bumbarger, Radiology Service, Walter Reed National Military Medical Center, Bethesda, MD.)