

Selected articles published by the Complement and Diseases team for the last 5 years

2014

1. Marinozzi MC, Vergoz L, Rybkine T, et al. Complement Factor B Mutations in Atypical Hemolytic Uremic Syndrome--Disease-Relevant or Benign? *J Am Soc Nephrol.* 2014.
2. Rayes J, Roumenina LT, Dimitrov JD, et al. The interaction between factor H and VWF increases factor H cofactor activity and regulates VWF prothrombotic status. *Blood.* 2014;123:121-125.
3. Dragon-Durey MA, Blanc C, Roumenina LT, et al. Anti-factor H autoantibodies assay. *Methods Mol Biol.* 2014;1100:249-256.
4. Roumenina LT, Roquigny R, Blanc C, et al. Functional evaluation of factor H genetic and acquired abnormalities: application for atypical hemolytic uremic syndrome (aHUS). *Methods Mol Biol.* 2014;1100:237-247.

2013

1. Lemaire* M, Fremeaux-Bacchi* V, Schaefer F, et al. Recessive mutations in DGKE cause atypical hemolytic-uremic syndrome. *Nat Genet.* 2013;45:531-536. * equal contribution
2. Fremeaux-Bacchi V, Fakhouri F, Garnier A, et al. Genetics and outcome of atypical hemolytic uremic syndrome: a nationwide French series comparing children and adults. *Clin J Am Soc Nephrol.* 2013;8:554-562.
3. Frimat M, Tabarin F, Dimitrov JD, et al. Complement activation by heme as a secondary hit for atypical hemolytic uremic syndrome. *Blood.* 2013;122:282-292.
4. Loupy A, Lefaucheur C, Vernerey D, et al. Complement-binding anti-HLA antibodies and kidney-allograft survival. *N Engl J Med.* 2013;369:1215-1226.
5. Le Quintrec M, Zuber J, Moulin B, et al. Complement genes strongly predict recurrence and graft outcome in adult renal transplant recipients with atypical hemolytic and uremic syndrome. *Am J Transplant.* 2013;13:663-675.
6. Hebecker M, Alba-Dominguez M, Roumenina LT, et al. An engineered construct combining complement regulatory and surface-recognition domains represents a minimal-size functional factor H. *J Immunol.* 2013;191:912-921.
7. Roumenina LT, Zuber J, Fremeaux-Bacchi V. Physiological and therapeutic complement regulators in kidney transplantation. *Curr Opin Organ Transplant.* 2013;18:421-429.
8. Nicolas C, Vuiblet V, Baudouin V, et al. C3 nephritic factor associated with C3 glomerulopathy in children. *Pediatr Nephrol.* 2013;29:85-94.
9. Skerka C, Chen Q, Fremeaux-Bacchi V, Roumenina LT. Complement factor H related proteins (CFHRs). *Mol Immunol.* 2013;56:170-180.
10. Fakhouri F, Fremeaux-Bacchi V. Thrombotic microangiopathy: eculizumab for atypical haemolytic uraemic syndrome: what next? *Nat Rev Nephrol.* 2013;9:495-496.
11. Dimitrov JD, Planchais C, Roumenina LT, Vassilev TL, Kaveri SV, Lacroix-Desmazes S. Antibody polyreactivity in health and disease: statu variabilis. *J Immunol.* 2013;191:993-999.

12. Bresin E, Rurali E, Caprioli J, et al. Combined complement gene mutations in atypical hemolytic uremic syndrome influence clinical phenotype. *J Am Soc Nephrol.* 2013;24:475-486.
13. Bekassy ZD, Kristoffersson AC, Cronqvist M, et al. Eculizumab in an anephric patient with atypical haemolytic uraemic syndrome and advanced vascular lesions. *Nephrol Dial Transplant.* 2013;28:2899-2907.

2012

1. Servais A, Noel LH, Roumenina LT, et al. Acquired and genetic complement abnormalities play a critical role in dense deposit disease and other C3 glomerulopathies. *Kidney Int.* 2012. 82:454-464.
2. Sartz L, Olin AI, Kristoffersson AC, et al. A novel C3 mutation causing increased formation of the C3 convertase in familial atypical hemolytic uremic syndrome. *J Immunol.* 2012;188:2030-2037.
3. Roumenina LT, Frimat M, Miller EC, et al. A prevalent C3 mutation in aHUS patients causes a direct C3 convertase gain-of-function. *Blood.* 2012. 119:4182-4191.
4. Malina M, Roumenina LT, Seeman T, et al. Genetics of hemolytic uremic syndromes. *Presse Med.* 2012;41:e105-114.
5. Ermini L, Goodship TH, Strain L, et al. Common genetic variants in complement genes other than CFH, CD46 and the CFHRs are not associated with aHUS. *Mol Immunol.* 2012;49:640-648.
6. El Karoui K, Hill GS, Karras A, et al. A clinicopathologic study of thrombotic microangiopathy in IgA nephropathy. *J Am Soc Nephrol.* 2012;23:137-148.
7. Blanc C, Roumenina LT, Ashraf Y, et al. Overall neutralization of complement factor H by autoantibodies in the acute phase of the autoimmune form of atypical hemolytic uremic syndrome. *J Immunol.* 2012;189:3528-3537.

2011 (n=20)

1. Servais A, Noel LH, Dragon-Durey MA, et al. Heterogeneous pattern of renal disease associated with homozygous factor H deficiency. *Hum Pathol.* 2011;42:1305-1311.
2. Salmon JE, Heuser C, Triebwasser M, et al. Mutations in complement regulatory proteins predispose to preeclampsia: a genetic analysis of the PROMISSE cohort. *PLoS Med.* 2011;8:e1001013.
3. Roumenina LT, Sene D, Radanova M, et al. Functional complement C1q abnormality leads to impaired immune complexes and apoptotic cell clearance. *J Immunol.* 2011;187:4369-4373.
4. Roumenina LT, Radanova M, Atanasov BP, et al. Heme interacts with c1q and inhibits the classical complement pathway. *J Biol Chem.* 2011;286:16459-16469.
5. Roumenina LT, Loirat C, Dragon-Durey MA, Halbwachs-Mecarelli L, Sautes-Fridman C, Fremeaux-Bacchi V. Alternative complement pathway assessment in patients with atypical HUS. *J Immunol Methods.* 2011;365:8-26.
6. Nilsson SC, Sim RB, Lea SM, Fremeaux-Bacchi V, Blom AM. Complement factor I in health and disease. *Mol Immunol.* 2011;48:1611-1620.
8. Loirat C, Fremeaux-Bacchi V. Atypical hemolytic uremic syndrome. *Orphanet J Rare Dis.* 2011;6:60.

9. Lapeyraque AL, Malina M, Fremeaux-Bacchi V, et al. Eculizumab in severe Shiga-toxin-associated HUS. *N Engl J Med*. 2011;364:2561-2563.
10. Fremeaux-Bacchi V, Fakhouri F, Roumenina L, Dragon-Durey MA, Loirat C. [Atypical hemolytic-uremic syndrome related to abnormalities within the complement system]. *Rev Med Interne*. 2011;32:232-240.
11. Camous L, Roumenina L, Bigot S, et al. Complement alternative pathway acts as a positive feedback amplification of neutrophil activation. *Blood*. 2011;117:1340-1349.
12. Bridoux F, Desport E, Fremeaux-Bacchi V, et al. Glomerulonephritis with isolated C3 deposits and monoclonal gammopathy: a fortuitous association? *Clin J Am Soc Nephrol*. 2011;6:2165-2174.
13. Brackman D, Sartz L, Leh S, et al. Thrombotic microangiopathy mimicking membranoproliferative glomerulonephritis. *Nephrol Dial Transplant*. 2011;26:3399-3403.

2010

1. Zuber J, Le Quintrec M, Sberro-Soussan R, Loirat C, Fremeaux-Bacchi V, Legendre C. New insights into postrenal transplant hemolytic uremic syndrome. *Nat Rev Nephrol*. 2010;7:23-35.
2. Sallee M, Daniel L, Piercecchi MD, et al. Myocardial infarction is a complication of factor H-associated atypical HUS. *Nephrol Dial Transplant*. 2010;25:2028-2032.
3. Rossi V, Bally I, Ancelet S, et al. Functional characterization of the recombinant human C1 inhibitor serpin domain: insights into heparin binding. *J Immunol*. 2010;184:4982-4989.
4. Nilsson SC, Kalchishkova N, Trouw LA, Fremeaux-Bacchi V, Villoutreix BO, Blom AM. Mutations in complement factor I as found in atypical hemolytic uremic syndrome lead to either altered secretion or altered function of factor I. *Eur J Immunol*. 2010;40:172-185.
5. Loirat C, Macher MA, Elmaleh-Berges M, et al. Non-atheromatous arterial stenoses in atypical haemolytic uraemic syndrome associated with complement dysregulation. *Nephrol Dial Transplant*. 2010;25:3421-3425.
6. Le Quintrec M, Roumenina L, Noris M, Fremeaux-Bacchi V. Atypical hemolytic uremic syndrome associated with mutations in complement regulator genes. *Semin Thromb Hemost*. 2010;36:641-652.
7. Koehl B, Boyer O, Biebuyck-Gouge N, et al. Neurological involvement in a child with atypical hemolytic uremic syndrome. *Pediatr Nephrol*. 2010;25:2539-2542.
8. Habibi I, Sfar I, Ben Alaya W, et al. Atypical hemolytic uremic syndrome and mutation analysis of factor H gene in two Tunisian families. *Int J Nephrol Renovasc Dis*. 2010;3:85-92.
9. Gale DP, de Jorge EG, Cook HT, et al. Identification of a mutation in complement factor H-related protein 5 in patients of Cypriot origin with glomerulonephritis. *Lancet*. 2010;376:794-801.
10. Fakhouri F, Roumenina L, Provot F, et al. Pregnancy-associated hemolytic uremic syndrome revisited in the era of complement gene mutations. *J Am Soc Nephrol*. 2010;21:859-867.
11. Fakhouri F, Fremeaux-Bacchi V, Noel LH, Cook HT, Pickering MC. C3 glomerulopathy: a new classification. *Nat Rev Nephrol*. 2010;6:494-499.
12. Dragon-Durey MA, Sethi SK, Bagga A, et al. Clinical features of anti-factor H autoantibody-associated hemolytic uremic syndrome. *J Am Soc Nephrol*. 2010;21:2180-2187.

13. Dragon-Durey MA, Blanc C, Garnier A, Hofer J, Sethi SK, Zimmerhackl LB. Anti-factor H autoantibody-associated hemolytic uremic syndrome: review of literature of the autoimmune form of HUS. *Semin Thromb Hemost.* 2010;36:633-640.
14. Dimitrov JD, Roumenina LT, Plantier JL, et al. A human FVIII inhibitor modulates FVIII surface electrostatics at a VWF-binding site distant from its epitope. *J Thromb Haemost.* 2010;8:1524-1531.
15. Chatelet V, Lobbedez T, Fremeaux-Bacchi V, Ficheux M, Ryckelynck JP, Hurault de Ligny B. Eculizumab: safety and efficacy after 17 months of treatment in a renal transplant patient with recurrent atypical hemolytic-uremic syndrome: case report. *Transplant Proc.* 2010;42:4353-4355.
16. Bienaime F, Dragon-Durey MA, Regnier CH, et al. Mutations in components of complement influence the outcome of Factor I-associated atypical hemolytic uremic syndrome. *Kidney Int.* 2010;77:339-349.

2009

1. Skerka C, Jozsi M, Zipfel PF, Dragon-Durey MA, Fremeaux-Bacchi V. Autoantibodies in haemolytic uraemic syndrome (HUS). *Thromb Haemost.* 2009;101:227-232.
2. Saland JM, Ruggenenti P, Remuzzi G. Liver-kidney transplantation to cure atypical hemolytic uremic syndrome. *J Am Soc Nephrol.* 2009;20:940-949.
3. Roumenina LT, Jablonski M, Hue C, et al. Hyperfunctional C3 convertase leads to complement deposition on endothelial cells and contributes to atypical hemolytic uremic syndrome. *Blood.* 2009;114:2837-2845.
4. Oberic L, Buffet M, Schwarzingler M, et al. Cancer awareness in atypical thrombotic microangiopathies. *Oncologist.* 2009;14:769-779.
5. Mache CJ, Acham-Roschitz B, Fremeaux-Bacchi V, et al. Complement inhibitor eculizumab in atypical hemolytic uremic syndrome. *Clin J Am Soc Nephrol.* 2009;4:1312-1316.
6. Licht C, Fremeaux-Bacchi V. Hereditary and acquired complement dysregulation in membranoproliferative glomerulonephritis. *Thromb Haemost.* 2009;101:271-278.
7. Lhotta K, Janecke AR, Scheiring J, et al. A large family with a gain-of-function mutation of complement C3 predisposing to atypical hemolytic uremic syndrome, microhematuria, hypertension and chronic renal failure. *Clin J Am Soc Nephrol.* 2009;4:1356-1362.
8. Lee BH, Kwak SH, Shin JI, et al. Atypical hemolytic uremic syndrome associated with complement factor H autoantibodies and CFHR1/CFHR3 deficiency. *Pediatr Res.* 2009;66:336-340.
9. Le Quintrec M, Zuber J, Noel LH, et al. Anti-Factor H autoantibodies in a fifth renal transplant recipient with atypical hemolytic and uremic syndrome. *Am J Transplant.* 2009;9:1223-1229.
10. Kwon T, Belot A, Ranchin B, et al. Varicella as a trigger of atypical haemolytic uraemic syndrome associated with complement dysfunction: two cases. *Nephrol Dial Transplant.* 2009;24:2752-2754.
11. Hirt-Minkowski P, Schaub S, Mayr M, et al. Haemolytic uraemic syndrome caused by factor H mutation: is single kidney transplantation under intensive plasmatherapy an option? *Nephrol Dial Transplant.* 2009;24:3548-3551.

12. Fuchs A, Atkinson JP, Fremeaux-Bacchi V, Kemper C. CD46-induced human Treg enhance B-cell responses. *Eur J Immunol.* 2009;39:3097-3109.
13. Dragon-Durey MA, Blanc C, Marliot F, et al. The high frequency of complement factor H related CFHR1 gene deletion is restricted to specific subgroups of patients with atypical haemolytic uraemic syndrome. *J Med Genet.* 2009;46:447-450.
14. Dimitrov JD, Roumenina LT, Andre S, et al. Kinetics and thermodynamics of interaction of coagulation factor VIII with a pathogenic human antibody. *Mol Immunol.* 2009;47:290-297.
15. Chatelet V, Fremeaux-Bacchi V, Lobbedez T, Ficheux M, Hurault de Ligny B. Safety and long-term efficacy of eculizumab in a renal transplant patient with recurrent atypical hemolytic-uremic syndrome. *Am J Transplant.* 2009;9:2644-2645.