

during CRRT. Dabigatran plasma clearance varied between 91 and 169 mL/min during IHD and remained constant during treatments. A 33% rebound in dabigatran levels occurred in one patient after IHD but incomplete data were available in others; rebound was not observed post-CRRT. Prolonged IHD (8–18 h, continuously) appeared sufficient to render dabigatran subtherapeutic, including the case requiring urgent surgery; four hours was insufficient in one case. Of the four patients with bleeding, one expired shortly after IHD.

Conclusions: IHD enhanced dabigatran elimination more than CRRT and complications were not observed. Dialysis decisions, including modality and duration must be individualised based on a risk-benefit assessment.

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RETINAL ABNORMALITIES IN THE C3 NEPHROPATHIES AND THE RISK OF VISUAL IMPAIRMENT

D COLVILLE¹, L AMOS¹, P HUGHES³, K NICHOLLS³, H MACK², RCA SYMONS^{1,2}, JD SMITH¹, J SAVIGE¹

¹The University of Melbourne Department of Medicine, Melbourne Health and Northern Health, Melbourne; ²Eye Surgery Associates, East Melbourne;

³Department of Nephrology, Royal Melbourne Hospital, Melbourne, Australia

Aims: To determine retinal abnormalities in 8 affected individuals from 7 families with DDD and two unrelated individuals with atypical HUS.

Background: The C3 nephropathies comprise dense deposit disease (DDD, formerly known as membranoproliferative glomerulonephritis type II), C3 glomerulonephritis and atypical haemolytic uraemic syndrome. All these diseases may be associated with mutations in the Complement Factor H gene (CFH), as well as other genes. Retinal drusen have been described in DDD, and we describe here retinal abnormalities in DDD and atypical HUS.

Methods: All individuals with DDD were examined by an ophthalmologist, and together with the patients with atypical HUS underwent retinal imaging and optical coherence tomography (OCT).

Results: Ocular symptoms in patients with DDD included impaired night vision, which was common and occurred early, and impaired peripheral vision. Individuals with DDD had multiple small basal laminar drusen (<125 µm), large soft drusen, or both. Drusen were first seen in adolescence, with pigmentation and haemorrhage occurring later. Optical coherence tomography (OCT) demonstrated the drusen location beneath the retinal pigment epithelium, secondary atrophy of the neuroretina in about half the individuals, and, in one, the further complications of sub-retinal choroidal neovascular membranes, pigment epithelial detachment and atypical serous retinopathy.

One of the two individuals with atypical HUS had large soft macular drusen (> 125 µm) identical to those seen in DDD.

Conclusions: Individuals with C3 nephropathy should be assessed ophthalmologically for macular disease at diagnosis, and monitored annually thereafter. They should be advised to present immediately if their vision deteriorates. Laser therapy prevents some retinal complications, and biological treatments offer further hope.

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ECULIZUMAB IS EFFECTIVE THERAPY FOR ATYPICAL HAEMOLYTIC URAEMIC SYNDROME (aHUS): A CASE SERIES OF AUSTRALIAN PATIENTS

A MALLET^{1,2}, S CAMPBELL^{2,3}, C VAN EPS^{2,3}, C HAWLEY^{2,3}, J BURKE^{2,3}, P HUGHES⁴, J KAUSMAN⁵, I HEWITT⁶, A PARNHAM⁷, J SZER⁴, A TUCKFIELD⁴, S PAYNE⁸, J YOUNG⁸, N ISBEL^{2,3}

¹Department of Renal Medicine, Royal Brisbane and Women's Hospital, Brisbane, Qld;

²The University of Queensland, Brisbane, Qld; ³Department of Nephrology, Princess Alexandra Hospital, Brisbane, Qld;

⁴Departments of Nephrology and Clinical Haematology, Royal Melbourne Hospital, Melbourne, Victoria;

⁵Department of Nephrology, Royal Children's Hospital, Melbourne, Victoria;

⁶Department of Nephrology, Princess Margaret Hospital for Children, Perth, WA;

⁷Department of Renal Medicine, Gold Coast University Hospital, Southport, Qld;

⁸Alexion Pharmaceuticals, Australia

Aim: To report on Australian patients with aHUS who have received the complement inhibitor, eculizumab, on compassionate grounds.

Background: aHUS is an ultra-rare, genetic, life-threatening disease associated with high rates of end-stage kidney disease and premature death. Chronic, uncontrolled complement activation causes systemic thrombotic microangiopathy (TMA), acute kidney injury and multi-organ system damage. Prior to the advent of eculizumab, 33–40% of patients with aHUS died or reached ESKD with their first manifestation of the disease.

Methods: Patients with a clinical diagnosis of aHUS who have received compassionate eculizumab therapy have been included in this case series.

Results: 10 patients with aHUS began treatment with eculizumab between 2010 and 2013. 80% of patients were female with presentation at 7 months to 40 years of age. The median duration of treatment was 12 months (range 4–42 months). All patients had haematological evidence of TMA and organ damage, had ADAMTS-13 activity of >5% and were negative for STEC. Five patients demonstrated multiple progressive extra-renal complications including severe hypertension, cardiomyopathy and neurological complications including headaches, fatigue, drowsiness, blurred vision, poor balance and seizures.

Eculizumab was well tolerated and all patients remain on drug with median follow-up of 18 months. All 9 surviving patients had a rapid haematological response with resolution of TMA and discontinued plasma exchange/infusions (PE/PI). Five patients (50%) were dialysis-dependent at initiation of eculizumab. Treatment with eculizumab was able to eliminate dialysis in 4 of these 5 (80%) patients. All patients experienced improvement or stabilisation of extra-renal manifestations.

Conclusions: aHUS is a severe multi-system disease and eculizumab treatment is a promising therapy that allows for preservation of renal function and elimination of dialysis and PE/PI.

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FOCAL SEGMENTAL GLOMERULOSCLEROSIS – PRESENTING FEATURES, TREATMENT AND OUTCOMES IN FAR NORTH QUEENSLAND ADULTS

AM GREENWOOD¹, R GUNNARSSON¹, RA BAER²

¹School of Medicine and Dentistry, James Cook University, Cairns, QLD;

²Department of Renal Medicine, Cairns Hospital, Cairns, QLD, Australia

Aim: To describe the clinical features, treatment and outcomes in adult patients with Focal Segmental Glomerulosclerosis (FSGS) presenting in Far North Queensland.

Background: FSGS is a common cause of end stage kidney disease (ESKD) understudied within the Australian population. The incidence of ESKD attributed to FSGS is increasing worldwide.

Methods: A retrospective review on 47 patients with biopsy confirmed FSGS between January 1997 and December 2013 at Cairns Hospital renal unit. Patients were located through the local biopsy database of 790 biopsies. Presenting features, secondary causes, prognostic factors, treatment modality and outcomes (doubling of serum creatinine, ESKD, death) were examined.

Results: Patients were followed on average for 4.8 years (range 2 months to 16 years). The mean age was 45 years with 50% of Aboriginal and Torres Strait Islander origin. Secondary causes included obesity (45%), hepatitis (6.8%) and additional glomerulonephritis (27%). FSGS not otherwise specified (NOS) was the most common variant (83%). Renin angiotensin system inhibitors and immunosuppression were commonly given (70% and 29% respectively). Of those given steroids, 45% responded and the remainder had an equal distribution of dependence or resistance. Eight patients died (17%), 14 doubled their serum creatinine (30%) and 13 progressed to ESKD (30%) within an average of 4.3 years. The number of patients was too few to make any statistically significant conclusions regarding prognostic factors.

Conclusions: FSGS holds serious implications for patients with 40% progressing to the combined outcome. The Aboriginal and Torres Strait Islander community is over-represented in this population. Rates of immunosuppression treatment suggest either a significant proportion of secondary FSGS or under-treatment of primary FSGS in the region. Detailed description of prognostic factors remains to be clarified.

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CORRELATION BETWEEN IMMUNOGLOBULIN G4 (IgG4) GLOMERULAR STAINING AND ANTI-PHOSPHOLIPASE A2 RECEPTOR ANTIBODY (PLA2R) TESTING IN MEMBRANOUS NEPHROPATHY

V RAGHUNATH¹, M SURANYI¹, J QUIN², P LEHMANN², J YONG³, A MAKKRIS¹

¹Department of Nephrology; ²Department of Immunology; ³Department of Pathology, Liverpool Hospital, Liverpool, NSW, Australia

Aim: To investigate the correlation of PLA2R antibody with clinical, biochemical and histological markers of membranous nephropathy.