A44
Long-term outcomes of patients with reversible cerebral vasocostriction syndrome (RCVS)

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Introduction.— RCVS is characterized by acute onset of severe headaches, with or without neurologic deficit with evidence of reversible cerebral vasocostriction [1]. Natural history and long-term outcome of RCVS has not been thoroughly investigated.

Methods.— Prospective cohort analysis of patients recruited from the RCVS registry at the Cleveland Clinic, was conducted. Validated questionnaires were mailed to the patients. The forms included: Headache screening form, Headache Impact Test-6 (HIT-6), Migraine Disability Assessment Test (MDAS), Barthel index (BI), Patient Health Questionnaire (PHQ-9) and EuroQol (EQ-5D-5L).

Results.— From a total of 57 patients, three refused, 26 were lost to follow-up, eight never replied and 20 participated. Median follow-up time was 91.5 months (range 10-254). Of the 20 patients (90% female), 19 (95%) presented with thunderclap headache and had initial ischemic stroke or hemorrhage, almost all were independent with one having worsening. Headache impact on life measured by HIT-six showed that two (13%) patients had a severe impact (HIT score > 60). The mean MIDAS score was 10.67 and two (13%) patients had severe disabling headaches (MIDAS score > 21). Sixteen (94%) patients were independent by BI scores > 85. EQ-SD-SL measurements showed that 12 (71%) 14 (82%) and 12 (71%) patients had no problems with mobility, self-care and leisure respectively.

Discussion.— These data on long-term outcomes suggests that half of them will continue to have headache, although decreased in severity and frequency. Although close to three-quarter of patients suffered an initial ischemic stroke or hemorrhage, almost all were independent with little disability. Pain and anxiety however decreased the QoL.

Conclusion.— This is the first study to address the long-term outcomes of patients with RCVS. The study findings indicate that long term outcome of patients with RCVS is favorable.

Reference

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A45
Recognizing childhood inflammatory brain diseases in Canada

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Introduction.— Childhood inflammatory brain diseases are life-threatening diseases causing devastating brain damage in previously healthy children. Over the past decade recognition has increased. Since 2007, the BrainWorks network prospectively captures children with inflammatory brain diseases. The aim of the study was to determine the spectrum of inflammatory brain diseases in Canadian children and identify presenting features of distinct subtypes.

Methods.— Consecutive children enrolled into the BrainWorks study at Canadian centers from January 2007 until September 2012 were identified. Children were included, if predetermined information (demographics, diagnosis, clinical, laboratory, neuroimaging, brain biopsy) at the baseline visit were complete.

Results.— In total 247 children were included; 132 boys, 115 girls mean age 9.3 yrs. The most common inflammatory brain diseases were; primary CNS vasculitis 90 non-progressive large vessel vasculitis (63 boys 27 girls mean age 8.2 yrs), 57 small vessel vasculitis (19 boys 38 girls mean age 11 yrs) 25 progressive large vessel vasculitis (21 boys four girls mean age 10.3 yrs); anti-NMDAR-encephalitis in 25 children (seven boys 18 girls mean age 9.9 yrs), other neuronal antibody mediated diseases six children; secondary CNS vasculitis 29 children, 13 associated with infection, ten underlying rheumatic disease, six systemic vasculitis. At presentation focal deficits were most common in large vessel CNS vasculitis, and diffuse deficits in small vessel CNS vasculitis and NMDAR encephalitis (61% and 72%). Seizures were seen more frequently in NMDAR encephalitis and small vessel CNS vasculitis (80% and 61%).

Conclusion.— Inflammatory brain diseases are increasingly diagnosed in Canadian children of all ages. In this study the majority presented with primary inflammatory CNS diseases. Vasculitis is the most common childhood inflammatory brain disease, however NMDAR encephalitis is increasingly recognized and has to be considered given the widely overlapping clinical presentation.

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A46
Factors predictive of prognosis in renal AAV – A study of 104 patients in a single UK centre

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Introduction.— The identification of predictive factors for renal and patient outcomes in AAV could guide therapy, which itself is toxic, and results in 1/3 of patient deaths within a year of diagnosis [1]. Our aim was to identify potential prognostic factors in patients treated at our centre.

Patients.— AAV patients were identified with at least ten glomerulii on biopsy and 1 year’s follow-up. Renal biopsies were classified and clinical data retrospectively reviewed. Patients were divided by histological classification according to the Berden classification [2], by degree of tubular atrophy (TA) (< 20%, 20-50% and > 50%), by ANCA subtype, P or C, and by starting eGFR (< 15, 15–60 and > 60). The primary outcome was a combined endpoint of renal and patient death.

Results.— One hundred and four patients were included, median age 62 yrs and 58% male. There were equal proportions (47%) of P and C-ANCA +ve patients. Twenty-three biopsies were classified as focal, 26 crescentic, 48 mixed and seven sclerotic. eGFR differed across the four histological classes at 1 year (P < 0.0001). Overall outcomes were best in the focal and worse in the sclerotic groups (figure 1). 9% normal glomerulii, degree of TA and starting eGFR were independent predictors
of outcome. C-ANCA positive patients had better outcome than P-ANCA positive.

**Discussion.**– There was a significant difference in outcomes between patients in the focal and sclerotic group at 5 yrs. Previous validation studies [2] have differed with regard to outcomes in the crescentic and mixed groups, which in our study were similar. The greatest improvement in eGFR at 1 and 5 yrs was in the crescentic class. Percentage normal glomeruli, degree TA and starting eGFR were previously correlated with outcome and our study confirmed these findings.

**Conclusion.**– This large study provides further validation of the prognostic use of Berden’s classification of renal AAV, and in addition highlights the importance of proportion of normal glomeruli, TA, starting eGFR and ANCA type as prognostic factors.

**References**


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**A47 Prognosis of patients with ANCA-associated glomerulonephritis (GN) presenting with severe renal failure**


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**Introduction**– Immunosuppressive treatment is currently offered to all patients with ANCA-GN presenting with severe renal failure in hope of restoring renal function. We evaluated prognostic factors impacting renal and patient survival for such patients.

**Patients**– Patients with ANCA-GN, eGFR < 15 mL/min/1.73 m² and treated with immunosuppressive therapy were included. Cumulative renal and patient survival rates were calculated with competing risk adjustment. Clinical and histologic characteristics were evaluated to determine prognostic factors. Histologic activity score (0–24 points for six items) and chronicity score (0–16 points: interstitial fibrosis, tubular atrophy, glomerular and crescentic sclerosis) were used. Treatment response was defined as patient survival with eGFR > 20 mL/min.

Multivariate analyses were performed with a Cox-proportional hazard model for long-term outcomes and logistic regression model for treatment response.

**Results**– One hundred and fifty-five patients were included: median age 67 years (IQR [53–74]); 56% male; 88% White; 56% MPO-ANCA; median eGFR 7 [5–9] mL/min/1.73 m²; 87% received cyclophosphamide, 28% plasmapheresis. By 4 months, 51% of patients attained treatment response, 35% remained on dialysis and 14% died. Renal and patient survival rates were 74% & 81%, and 68% & 67% at 1 and 5 years respectively. The probability of treatment response at 4 months was increased with eGFR > 10 (OR, 2.78 [95% CI, 1.09–7.12] vs. eGFR < 10) and low chronicity score (OR, 1.13 [1.02–1.22] per unit decrease). Treatment response at 4 months and cyclophosphamide use were independent predictors of long-term renal/patient survival, adjusting for baseline eGFR, % normal glomeruli and chronicity score. The frequency of response beyond 4 months was only 3.6%.

**Conclusion**– Although baseline eGFR < 10 and high chronicity score are associated with lower response rate, no “futility” threshold was identified. Conversely, continued immunosuppressive therapy beyond 4 months is very unlikely to benefit patients who remain dialysis dependent.

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**A48 Diffuse alveolar haemorrhage (DAH) in levamisole-adultered cocaine abuser resolved by rFVIIa infusion**

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**Introduction**– Vasculitis induced by levamisole-adultered cocaine is a new entity described since 2008. It mainly involves skin with necrotic erythematous purpuric lesions located at nose, cheeks, earlobes and extremities. Another main complication is agranulocytosis. Fever, malaise and artho-myalgias are usually present. It is associated with presence of autoantibodies: ANA, ANCA with predominant anti-MPO specificity, antiphospholipids antibodies and low C3 values. Differently from ANCA-Associated vasculitis, kidneys and lung are rarely involved. We report a case with severe DAH in cocaine abuser. Neither steroid pulses nor plasma-exchange were able to stop lung bleeding, which only resolved after rFVIIa administration.

**Results**– A 37-yr cocaine abuser male was admitted with 3 days history of malaise, thoracic pain, diffuse arthralgias. Physical examination revealed purpura and necrosis on lower extremities and earlobes, perforated septum with active crushing. CT scan showed right interstitial diffuse alveolar infiltrates, suggestive for DAH. Bronchoscopy confirmed DAH by identification of ongoing bleeding from the right bronchial tree; no positive bacterial, fungal or viral organisms were identified. Bronchial biopsy proved vasculitis. Pt rapidly developed haemoptysis and respiratory insufficiency. A chest X-ray demonstrated bilateral rapidly worsening air space disease. Urinalysis revealed ematuria and proteinuria, sCreatinine was normal. Laboratory exams showed ANCA anti-MPO at 67.9 IU/mL (nv < 19.99), antiphospholipids antibodies were negative. IV antibiotics, high doses methylprednisolone (1 g iv), transfusion support and plasmapheresis were administered. On day 7 the bleeding was still ongoing, thus rFVIIa was given (7 mg i.v.), in order to obtain