**A44**  
**Long-term outcomes of patients with reversible cerebral vasocostruction 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 vasocostruction [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 (MIDAS), 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 throbbing headache and had ischemic stroke (50%), subarachnoid (45%) or intracerebral (15%) hemorrhage. Eleven (55%) patients continued to have headache, but majority (91%) reported improvement in character with only 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-5L 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 4 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 glomeruli on biopsy and 1 year’s follow-up. Renal biopsies were classified and clinical data retrospectively reviewed. Patients were divided by histological class 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). % normal glomeruli, degree of TA and starting eGFR were independent predictors