by measuring nasal nitric oxide (nNO), a screening test for Primary Ciliary Dyskinesia (PCD). *Study design:* We measured nNO levels of 26 children with congenital midline CNS defects. We evaluated the effect of age, gender, and anomaly (brain, spinal cord, or combined) on measurements. We compared our results to the previously established normal range (153.6-509.9 nL/min), and to the cutoff for PCD (77 nL/min). *Results:* The range for nNO in our cohort was 56.5-334.7 nL/min, with age, gender, and anomaly not having a significant effect. The overall mean, 217.7 nL/min, was significantly lower than that of normal children, 314.51 nL/min ($p<0.01$). Four subjects (15.4%) had nNO levels below the lower end of normal, with two (7.7%) having values fitting the cutoff for PCD. *Conclusions:* We report an association between ciliary dysfunction and isolated midline neuroanatomical defects, not in context of any known syndrome. This suggests that genes causing isolated CNS defects, may be implied in the function of cilia. Longitudinal studies are required to investigate whether children with abnormal measurements suffer from any respiratory sequelae.

P.005

Utilization of transition care management plans to facilitate transition of adolescents with epilepsy into the adult healthcare system

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Background: For adolescents with epilepsy, there is often a poor system in place to meet their individualized transition needs. Our objectives were 1) to develop epilepsy-specific transition care management plans (TCMPs) to ensure access, and attachment to adult healthcare providers, and 2) to identify strategies for providing support during the transition period, including through the development of physician and patient (or caregiver) navigated web-based tools, resources and recommendations for health system improvements. *Methods:* Physicians and nurses with expertise in areas including adult and pediatric epilepsy, family medicine, psychiatry, and varied allied health professionals were engaged to generate epilepsy-related TCMPs. *Results:* Through an iterative process spanning the course of over a year, TCMPs were developed to cover areas including: treatment responsive and resistant epilepsy, ketogenic diet, epilepsy surgery, women's issues, mental health, and psychosocial aspects of epilepsy. The TCMPs referenced established guidelines and best practices in the literature wherever possible. Caregiver roles and responsibilities were outlined, remaining cognoscent of available provincial resources. *Conclusions:* Epilepsy specific TCMPs can be developed through a collaborative approach between pediatric and adult healthcare providers, easing the patient experience, creating educated accountability, and providing a forum to identify and address gaps of care in adolescents with epilepsy.

Child Neurology (General Pediatric Neurology)

P.006

Increased healthcare services utilization in the tuberous sclerosis complex population in Quebec

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Background: Tuberous sclerosis complex (TSC) is a neurocutaneous syndrome that can present with many disabling neurological symptoms, the most common being seizures. Although it is a chronic systemic syndrome, healthcare utilization and long-term outcome of subjects with TSC are not well defined. The goal of this study was to evaluate the direct cost and long-term outcome of TSC compared to other forms of epilepsy and healthy controls. *Methods:* Our provincial health care database was interrogated to determine use of medical services by patients with TSC, epilepsy and healthy controls from 1996-2011. Data on demographics, outcomes and health care utilization were analyzed. *Results:* 1004 TSC, 41,934 with epilepsy and 41,934 controls were identified. The prevalence of TSC was 1/7,872 compared to 1/189 for epilepsy. TSC experienced more hospitalizations, medical visits and prescription drug use, resulting in higher total health care costs. Their most common admission diagnosis was seizures and age at death was significantly lower: 61,3 years old for TSC vs 69,6 and 76,6 years old for epilepsy and controls, ($p<0,001$). *Conclusions:* TSC subjects have a significantly higher burden of disease than other subjects with epilepsy. These results stress the need for specialized services in this population through the lifespan.

P.007

Topographical orientation as a model of plasticity in children with perinatal stroke

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Background: Children with perinatal stroke go on to develop most cognitive skills (e.g. language) due to brain plasticity; however, their performance is usually poor when compared to age-matched controls, indicating a reduced potential compared to uninjured children. To date, how plasticity after early injury affects the development of complex cognitive skills remains uncertain. Here, we use topographical orientation, which relies on integration of several cognitive processes underlain by widespread neural networks, as a model to test plasticity in complex behaviour. *Methods:* Children with perinatal stroke and age-matched controls were tested with a neuropsychological battery and a novel navigation task. In addition, for each patient, we obtained the most recent MRI scan to assess the effects of lesion characteristics on performance at the navigational task. *Results:* Children with history of injury performed worse than controls, and their scores were not different based on lesion's laterality, location or functional region affected. In particular, involvement of regions known to contribute to spatial orientation did not result in significantly decreased performance. *Conclusions:* As seen in other

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skills, orientation was preserved, but decreased when compared to age-matched controls. Given its cognitive and neural complexity, topographical orientation may be used as a model for network plasticity after early injury.

P.008

Cerebral proliferative angiopathy in a three-year-old

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Background: Cerebral proliferative angiopathy (CPA) is a rare vascular malformation with intervening normal brain tissue interspersed among abnormal vascular channels. There are 77 reported cases, the youngest being 9 years old, with persistent symptoms and recurring symptoms being rare. *Methods:* A three-year-old girl with CPA is described and compared to the literature. *Results:* A previously healthy girl with early left-handedness and a left forehead nevus flammeus presented with sudden onset of right arm and leg weakness, along with abrupt speech arrest and right homonymous hemianopia. Head CT Angiogram and MRI revealed an abnormal vascular network with densely packed, moderately enlarged vessels arising within the white matter with no dominant feeding vessel and both old and acute infarcts in the left hemisphere. Eye exam was unremarkable. The clinical and radiologic features were most consistent with a diagnosis of CPA. Her visual deficits and motor symptoms persisted, and she had a recurrent event shortly after. *Conclusions:* This is the youngest reported case of CPA, with novel features including radiologic evidence of previous infarcts, clinical recurrence of symptoms, and permanent deficits. This case demonstrates the need for further research into the surveillance and management of this rare entity, possibly unique in young children

P.009

Successful treatment of paroxysmal tonic upgaze with low dose Gravol ®

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Background: We present a case of paroxysmal tonic upgaze (PTU) of infancy treated with a daily low dose of Gravol ® to improve symptoms. *Method:* Case report *Results:* A one year-old boy presented with episodes of sustained conjugate upgaze that persisted for 30 to 45 minutes, varied in severity, and occurred with increasing frequency over the past two months. The episodes were worse when fatigued and were relieved by sleep. Pregnancy, delivery, and development were normal. Neurological examination between episodes was normal, as were EEG, brain MRI, and blood analysis. CSF neurotransmitter analysis showed serotonin and dopamine metabolites at lower levels of normal. The patient was diagnosed with paroxysmal tonic upgaze of infancy and was treated with 12.5 mg of Gravol ® daily with complete cessation of episodes. *Conclusions:* Paroxysmal tonic upgaze (PTU) of infancy is a disorder seen in infants where the eyes are forcibly deviated upwards for minutes to hours at a time. PTU often resolves spontaneously over several months, however episodes are extremely debilitating. Currently, treatments with levodopa have been tried with some success. Via its anticholinergic effects, Gravol may be a novel therapeutic option for PTU, negating the need to use serotonergic medications.

Child Neurology (Neurocritical Care/Neuro Trauma)

P.010

Bacterial meningitis secondary to an intranasal encephalocele presenting as unilateral facial nerve palsy

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Background: Focal neurological deficits occur in approximately 15% of children with bacterial meningitis. However, cranial nerve involvement such as facial-nerve palsy is uncommon in non-tuberculous bacterial meningitis. *Methods:* Case Report. Review of the literature was conducted on Pubmed for the search terms: facial nerve palsy and meningitis. *Results:* We present the case of a 4-year old right-handed girl who presented with a new onset unilateral facial nerve palsy preceded by 5-day history of fever and headaches. The patient had meningeal signs and was identified to have Streptococcal Meningitis. MRI of the brain showed a large previously undiagnosed intranasal encephalocele. The facial palsy resolved within 7 days of antibiotic treatment. *Conclusions:* Our case represents an unusual combination of facial nerve palsy in context of Streptococcal Meningitis secondary to intranasal encephalocele.

Multidisciplinary

P.011

The cerebrovascular response to ketamine: a systematic review of the animal and human literature

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Introduction: Ketamine, an N-methyl D-aspartate (NMDA) receptor antagonist, carries potential benefit in patients with neurological illness. The cerebrovascular/cerebral blood flow (CBF) response to ketamine has been poorly outlined in the literature. *Methods:* We performed a systematic review of the literature on the cerebrovascular/CBF effects of ketamine in both animal and human subjects. *Results:* We identified 38 animal studies, and 20 human studies. Within the animal studies, a variety of different models were utilized with the majority focusing changes in global CBF or regional cerebral blood flow (rCBF). Overall, ketamine led to an increase in either global CBF or rCBF, with a vasodilatory effect in medium cerebral vessels. With the human studies a total of 379 patients, 107 of which were control subjects, were studied. Most studies focused on either 131Xe CT or PET imaging with ketamine administration. There was a trend to an increase in global CBF and rCBF with ketamine administration. *Conclusions:* Animal models indicate an increase in global CBF and rCBF with ketamine administration. Human studies display

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