

www.cpsp.cps.ca/surveillance. **Results:** Eleven cases were reported in 2020. Five (45%) cases were reported from Ontario and the remaining cases were reported from Atlantic Canada and Western Canada. Their median age was 12 months (IQR 6–21); 64% were male. The most common presenting symptoms were delayed motor milestone and hypotonia in 7 (64%) cases. On average, the diagnosis was delayed after the onset of symptoms by three months for SMA Type 1, by eight months for Type 2, and by 18 months for Type 3. Eight (73%) cases received nusinersen as their first disease-modifying treatment. **Conclusions:** Early recognition and newborn screening are essential to reduce diagnostic delay and enable timely treatment of SMA. Other data sources including the Canadian Neuromuscular Disease Registry and molecular genetic laboratories will be used to estimate the annual incidence of pediatric SMA in Canada.

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A Population-based Study of the Epidemiology, Healthcare Resource Utilization and Costs of Duchenne Muscular Dystrophy in Alberta, Canada

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Background: Duchenne muscular dystrophy (DMD) is a severe progressive neuromuscular disease. This study aimed to estimate the prevalence, healthcare resource utilization (HRU), and medical costs of DMD in Alberta. **Methods:** This retrospective study linked provincial healthcare administrative data to identify patients with DMD utilizing a modified diagnostic code algorithm, including males <30 years of age. Five-year (April 2012 to March 2017) prevalence estimates were calculated and all-cause direct HRU and costs were examined in the first-year post-diagnosis. **Results:** Overall, 111 patients (median age: 12.0 years (IQR 4.7-18.3)) with DMD were identified. The estimated five-year period prevalence was 35.72 (95% CI 31.88-39.91) per 100,000 persons. All-cause HRU in the first-year post-diagnosis included a mean (SD) of 0.48 (1.19) hospitalizations (length of stay: 9.37 days (36.47)), 3.96 (6.16) general practitioner visits, 28.52 (62.98) specialist visits, and 20.14 (16.49) ambulatory care visits. Mean (SD) all-cause direct costs were \$18,868 (\$29,206) CAD in the first-year post-diagnosis. **Conclusions:** Patients with DMD had multiple interactions with the healthcare system in the year following diagnosis, resulting in substantial direct medical costs. More effective treatment strategies are needed to improve health outcomes and reduce the burden of DMD.

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Benign tumors of peripheral nerves in children at a tertiary-care pediatric hospital

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Background: Tumors affecting peripheral nerves in children are rare. Accurate diagnosis ensures that management is appropriate and timely. **Methods:** We review the clinical presentation and utility of investigations of children with intrinsic tumors affecting peripheral nerves at the Children's Hospital of Eastern Ontario (CHEO). **Results:** From 2009-2019, 14 cases were identified. Mean age of symptom onset was 8.2 years (range 0.3 to 17.3 years). Presenting symptoms included painless muscle wasting (2/14), focal muscle weakness (7/14), contracture (1/14), pain (1/14) or a painless, palpable mass (3/14). MRI was useful at differentiating benign pediatric nerve tumors. Peripheral nerve lipomatosis demonstrated a classic "spaghetti string" appearance. Patients with perineurioma showed evidence of enhancing, nodular lesions while intraneural ganglionic cysts display cystic lesion within the nerve. Neurofibromas appear like a "bag of worms" while schwannomas are more eccentrically positioned around the nerve. Nerve conduction studies (NCS) or electromyography (EMG) were performed in 11/14 patients. Biopsies were performed in 9 patients and surgical management in 4 patients. **Conclusions:** The rare nature of peripheral nerve tumors in children can pose diagnostic challenges. NCS/EMG are important to assist with localization, and MRI important at distinguishing benign tumors. Key MRI, clinical and NCS features can guide management, potentially avoiding invasive procedures.

NEUROVASCULAR, STROKE AND NEUROINTERVENTIONAL

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Vessel Wall Imaging of Unusual Childhood Strokes: a Pediatric Case Series

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Background: MR-based vessel wall imaging (VWI) has gained influence in the clinical investigations, and management of pediatric strokes. Limitations still exist in interpreting it as a singular modality. **Methods:** We present 4 pediatric stroke cases

with VWI enhancement. **Results: Case 1.** 4-year old boy with sickle cell anemia, who developed encephalopathy during a hemolytic crisis. MR-VWI revealed bilateral extracranial internal carotid enhanced narrowing, deemed a secondary vasculopathy, with resolution upon follow-up.

Case 2. 16-year old male presented with left middle cerebral artery (MCA) infarction. VWI revealed left internal carotid terminus and proximal MCA enhancement. Conventional angiography showed abnormalities in mesenteric and hepatic arteries. Stability sustained on anticoagulation and immunosuppressive therapy.

Case 3. 10-year old girl, developed bilateral MCA infarctions with enhanced extracranial segments of both ICAs, and narrow PCAs, consistent with Moyamoya vasculopathy. Improved on combined immunosuppressive and anticoagulation therapy.

Case 4. 13-year old boy had an episode of right facial weakness, with a normal neurological exam; with enhancement and narrowing in the left extracranial ICA, likely an intramural hematoma from dissection. He responded to dual anticoagulation therapy. **Conclusions:** In conclusion, these cases illustrate similarities in vessel wall imaging abnormalities under different clinical contexts, with practical utility in longitudinal follow-up and prognostication.

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Cerebral Sinovenous Thrombosis in Preterm Infants

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Background: Neonatal cerebral sinovenous thrombosis (CSVT) can lead to severe brain injury and long-term neurodevelopmental impairments. Previous studies of neonatal CSVT have mainly included term infants. In this study, we examined the clinical and radiological features, treatment and outcome of CSVT in preterm infants. **Methods:** This was a retrospective cohort study of preterm infants born <37 weeks with radiologically confirmed CSVT. All MRI/MRV and CT/CTV scans were re-reviewed. Clinical and radiological data were analysed using descriptive statistics, ANOVA and chi-square tests. **Results:** A total of 26 preterm infants with CSVT were included. Of these, 65% were late preterm, 27% very preterm and 8% extreme preterm. Most were symptomatic (seizures 50%, abnormal exam 50%). Radiological features included transverse sinus (85%) and sagittal sinus thrombosis (42%), intraventricular hemorrhage (42%) and venous infarction (19%). Most preterm infants with CSVT (69%) were treated with anticoagulation. Anticoagulation was not associated with new or worsening intracranial hemorrhage. Outcome at follow-up ranged from no impairment (39%), mild impairment (19%), severe impairment (19%) and death (23%). **Conclusions:** Preterm infants with CSVT are often symptomatic and present with a distinct pattern of brain injury. Anticoagulation treatment of preterm CSVT appeared to be safe. Further studies and treatment guidelines for preterm CSVT are needed.

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Pediatric acute ischemic stroke protocols

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Background: Approximately 1,000 children present with AIS annually in North America. Most suffer from long-term disability. Childhood AIS is diagnosed after a median of 23 hours post-symptom onset, limiting thrombolytic treatment options that may improve outcomes. Pediatric stroke protocols decrease time to diagnosis. AIS treatment is not uniform across Canada, nor are pediatric stroke protocols standardized. **Methods:** We contacted neurologists at all 16 Canadian pediatric hospitals regarding their AIS management. **Results:** Response rate was 100%. Seven centers have an AIS protocol and two have a protocol under development. Seven centers do not have a protocol – two redirect patients to adult neurology, and five use a case-by-case approach for management. Analysis of the seven AIS protocols reveals differences: 1) IV-tPA dosage: age-dependent 0.75-0.9 mg/kg (n=1) versus age-independent 0.9 mg/kg (n=6), with maximum doses 75 mg (n=1) or 90 mg (n=6); 2) IV-tPA lower age cut-off: 2 years (n=4) versus 3, 4 or 10 years (n=1); 3) IV-tPA exclusion criteria: PedNIHSS score <4 (n=3), <5 (n=1), or <6 (n=3); 4) Pre-treatment neuroimaging: CT (n=3) versus MRI (n=4); 5) Intra-arterial tPA use (n=3). **Conclusions:** The seven Canadian pediatric AIS protocols show prominent differences. We plan a teleconference discussing a Canadian pediatric AIS consensus approach.

OTHER CHILD NEUROLOGY

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Curriculum mapping can facilitate transition to Competence by Design

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Background: Curriculum maps outline the content of an educational program identifying links between targeted outcomes, educational opportunities, and assessments. The transition to Competence by Design (CBD) in Canadian specialty residency programs requires thoughtful reorganization of educational programming. A curriculum map may assist with understanding the existing curriculum and thereby facilitate planning for CBD. **Methods:** A map of the pediatric neurology residency curriculum at the University of Calgary was constructed by linking objectives with related learning activities and assessments. Qualitative line-by-line analysis was then conducted to identify gaps in the existing curriculum. The map was used as a framework to plot CBD outcomes and curricular structure as these were established. **Results:** Generating the traditional curriculum map was time-consuming, requiring 48 hours. Careful review identified several objectives that did not link to formal learning activities or assessments. Many such gaps were recognized to link to non-clinical activities. Using the scaffold of the traditional curriculum reduced the