visit, 22/24 (92%) infants had achieved WHO motor milestones sitting without support and 8/16 (50%;2 *SMN2*, n=3/11;3 *SMN2*, n=5/5) on study >13 months achieved walking alone. AEs were reported in 24/25 (96%) infants; most 20/25 (80%) had AEs that were mild/moderate in severity; 9 had serious AEs. Four infants had an AE possibly related to study drug, which resolved despite continued treatment. No new safety concerns were identified. **Conclusions:** Nusinersen continued to benefit infants who initiated treatment in a presymptomatic stage of SMA.

Study Support: Biogen

B.06

Safety and efficacy of nusinersen in infants/children with spinal muscular atrophy (SMA): part 1 of the phase 2 EMBRACE study

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doi: 10.1017/cjn.2018.94

Background: EMBRACE (NCT02462759) Part 1 is a randomized, double-blind, sham-procedure controlled study assessing safety/tolerability of intrathecal nusinersen (12-mg equivalent dose) in symptomatic infants/children with SMA who were not eligible to participate in ENDEAR or CHERISH. **Methods:** Eligible participants had onset of SMA symptoms at ≤6 months with 3 *SMN2* copies; onset at ≤6 months, age >7 months and 2 copies; or onset at >6 months, age ≤18 months, and 2/3 copies. Safety/tolerability was the primary endpoint. Exploratory endpoints included Hammersmith Infant Neurological Examination Section 2 (HINE-2) motor milestone attainment, change in ventilator use, and growth. **Results:** EMBRACE Part 1 was terminated early based on positive results from ENDEAR. Safety/tolerability was similar to previous trials. More nusinersen-treated (11/14;79%) vs. sham–treated individuals (2/7;29%) were HINE-2 motor milestone responders. Between Day 183 and 302, mean (SD) hours of ventilator use changed by +1.236 (3.712) hours in nusinersen-treated (n=12) and +2.123 (3.023) hours in sham–treated individuals (n=7). Similar increases in weight and body length were observed in nusinersen-treated and sham–treated individuals by Day 183. **Conclusions:** In EMBRACE Part 1, nusinersen demonstrated a favorable benefit-risk profile. These results add to the aggregated efficacy, safety/tolerability data of nusinersen in SMA.

Study Supported by: Ionis and Biogen

B.07

Review of patients with Spinal Muscular Atrophy treated with Nusinersen in Ontario

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doi: 10.1017/cjn.2018.95

Background: Spinal Muscular Atrophy (SMA) is an autosomal recessive neurodegenerative disease. In June 2017, Health Canada approved Nusinersen, currently the only available drug for SMA. Since 2016, patients in Ontario have been treated clinically with Nusinersen through different access programs. **Methods:** Retrospective case series of patients with SMA treated clinically with Nusinersen in Ontario, describing clinical characteristics and logistics of intrathecal Nusinersen administration. **Results:** Twenty patients have been treated across four centres. To date, we have reviewed 8 cases at one centre (seven SMA Type I, one SMA Type II). Age at first dose ranged from 3-156 months and disease duration 9-166 months. Patients had received 4-7 doses at last evaluation. Three patients with scoliosis (2 with spinal rods) required fluoroscopy-guided radiologist administration, and 4 required general anesthesia. No complications/adverse events were reported. At last follow up, 5/8 families reported improved daily activities. Of 5 patients with baseline and follow up motor function testing, 3 demonstrated improved scores. One patient died due to respiratory decline at age 9 months, despite improved motor outcome scores. **Conclusions:** We describe the first Canadian post-marketing experience with Nusinersen. Timely dissemination of this information is needed to guide clinicians, hospital administrators, and policy-makers.

CNSS Chair's Select Abstracts

C.01

Endoscopic versus open microvascular decompression of trigeminal neuralgia: a systematic review and comparative meta-analysis

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doi: 10.1017/cjn.2018.96

Background: Microvascular decompression (MVD) is commonly used in the treatment of trigeminal neuralgia with positive clinical outcomes. Fully endoscopic microvascular decompression (E-MVD) has been proposed as a minimally invasive, effective alternative, but a comparative review of the two approaches in the literature has not been conducted. **Methods:** We performed a meta-analysis comparing patient outcome rates and complications for both techniques. From a pool of 1,039 studies, 22 articles were selected for review: 12 open MVD and 10 E-MVD. The total number of patients was 6,734. **Results:** Good pain relief was achieved in 81% of MVD and 88% of E-MVD patients, with a mean recurrence rate of 14% and 9% respectively. Average rates of complications in MVD versus E-MVD included facial paresis or weakness, 9%, 3%; hearing loss,

4%, 1%; cerebrospinal leak, both 3%; cerebellar damage and infection, 2%, <1%; and mortality <1%, 0% respectively. **Conclusions:** The reviewed literature revealed similar clinical outcomes with respect to pain relief for both approaches. Recurrence rate and incidence of complications, notably facial paresis and hearing loss were higher for MVD. We concluded that E-MVD appears to offer at least as good a surgical outcome as MVD, with possibly a shorter operative time, smaller craniectomy and lower recurrence rates.

C.02

Predictors of survival in a surgical series of Metastatic Spinal Cord Compression and a complete external validation of 8 models in a prospective multi-centre study

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doi: 10.1017/cjn.2018.97

Background: We aimed to identify preoperative predictors of survival in Metastatic Epidural Spinal Cord Compression (MESCC) patients surgically treated, examine how these predictors relate to eight prognostic models, and to perform the first full external validation of these models in accordance with the TRIPOD statement. **Methods:** 142 surgically treated MESCC patients were enrolled in a prospective, multicenter cohort study and followed for 12 months or until death. Cox proportional hazards (PH) regressions were used. Non-collinear predictors with <10% missing data, ≥10 events per stratum and p<0.05 in univariable analysis were tested through a backward stepwise selection. For the original and revised Tokuhashi, Tomita, modified Bauer, van der Linden, Bartels, OSRI, Bartels and Bollen, we examined calibration and discrimination; survival stratified by risk groups with the Kaplan-Meier method and log-rank test. **Results:** Primary tumor, organ metastasis and SF-36v2 PC were associated with survival in multivariable analysis; corrected discrimination was 0.68. These three predictors were common to most current prognostic models. However, calibration was poor overall while discrimation was possibly helpful. **Conclusions:** Primary tumor type (breast, prostate and thyroid), absence of organ metastasis, and a lower degree of physical disability are preoperative predictors of longer survival in surgical MESCC patients. Clinicians should use these 8 prognostic models with caution.

C.03

Surgical complications with and without image guidance: meta-analysis of Ommaya reservoir insertions

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doi: 10.1017/cjn.2018.98

Background: There remains an important role for consolidating evidence on the utility of image guidance (IG) in neurosurgery. In 1963, Ayub Ommaya proposed a surgical technique for the placement of a subcutaneous reservoir and pump to allow access to intraventricular cerebrospinal fluid. In this study, we sought to compile evidence from the literature about surgical outcome in ORI with and without IG. **Methods:** A systematic review was conducted in accordance with PRISMA guidelines. Overall surgical complication rate was considered a primary outcome and further classified into specific complication categories. **Results:** 40 studies were identified, including our own series, for a total of 1947 independent ORI procedures. Pooled rates of outcome for IG compared to non-IG were 6.0% versus 13.6% for overall complications; 2.0% versus 2.8% for catheter malfunction; 1.9% versus 2.3% for catheter malposition; 0.5% versus 4.0% for early infection; 4.3% versus 9.4% for any infection; and 0.4% versus 1.4% for mortality. **Conclusions:** We observed that IG ORI resulted in improved accuracy and decreased complications compared to non-IG. To our knowledge, this study comprises the largest observational analysis of operative outcomes demonstrating evidence for the utility of IG.

C.04

A systematic review and meta-analysis of 7551 patients with post-operative radiation for the management of functioning and non-functioning pituitary adenomas

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doi: 10.1017/cjn.2018.99

Background: Although surgery is the mainstay of treatment for most pituitary adenomas, post-operative radiotherapy has been shown to be of benefit in improving tumor control and recurrence-free survival. To understand potential side effects of radiotherapy we performed a systematic review and meta-analysis to determine the efficacy and safety of post-operative radiotherapy for pituitary adenoma. **Methods:** A systematic review was performed according to the Meta-analysis Of Observational Studies in Epidemiology (MOOSE) guidelines. We searched PubMed, MEDLINE and Cochrane databases with no language or publication date restrictions. Outcomes included 5- and 10-year progression-free survival and adverse events rates. **Results:** A total of 48 studies from 1986-2016 met the inclusion criteria, with 7551 cumulative patients. The cumulative 5- and 10-year s progression-free survival rates were 90.8% (95% CI 86-94%) and 88.6% (95% CI 81-93%), respectively. The overall adverse events rate was 8% (95% CI 5-12%). All outcomes were associated with significant heterogeneity ($I^2 \geq 70\%$). No differences in survival rates or adverse events in relation to study date, tumor pathology, radiosurgery system used or dose of radiation. **Conclusions:** Post-operative radiotherapy for pituitary adenomas is effective and safe. Because of the significant heterogeneity and lack of matched controls in the literature, optimum timing and dosage are still unclear. Further prospective studies are needed.