in CA2, 18 (10.1%) were in CA3/CA4, and 27 (15.2%) were in dentate gyrus. Along the longitudinal axis, hippocampal electrodes were most commonly implanted in the body (92; 51.7%) followed by the head (86; 48.3%). **Conclusions:** 7T MRI enables high-resolution anatomical imaging on the submillimeter scale in *in vivo* subjects. Here, we demonstrate the utility of 7T imaging for identifying the relative location of SEEG electrode implantations within hippocampal substructures for the invasive investigation of epilepsy.

P.064

Preoperative mapping using fMRI and DTI: a multimodal approach to assessing language dominance

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Background: Language mapping is a key goal in neurosurgical planning. With the discontinuation of the Wada test in Canada, neurosurgeons often rely on fMRI and intraoperative techniques for determining language lateralization. Recent studies have also evaluated the utility of diffusion tensor imaging (DTI) for preoperative language lateralization, but further research is needed to confirm its efficacy. We report a patient with a left frontal AVM. fMRI and DTI was used to localize language and motor functioning. **Methods:** The tasks included word reading, picture naming, pseudohomophones (e.g., dawg) and semantic questions. All fMRI analyses were performed using BrainVoyager. Tensors were tracked from 30-direction diffusion MR images using DSI-Studio. **Results:** The fMRI results revealed consistent Broca's and Wernicke's areas, confirming left hemisphere dominance. There was also a region of activation in the precentral gyrus near the surgical resection. The results were loaded onto the neuronavigation system to help determine safe surgical margins. The DTI results revealed that the left arcuate and uncinate fasciculus had three times more tracts than the right hemisphere, further supporting left hemisphere dominance. **Conclusions:** This case highlights the value of a combined, multimodal approach for preoperative language localization, which will further enhance surgical safety by helping preserve regions for essential brain functions.

P.066

Cortical autonomic patterns in Neurogenic Orthostatic Hypotension

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Background: Neurogenic orthostatic hypotension (NOH), defined as a drop in systolic blood pressure (SBP) ≥30mmHg on standing or head-up tilt, is associated with autonomic dysfunction. The cortical autonomic network (CAN) is a network of brain regions associated with autonomic function. Our aim was to investigate CAN activation patterns in NOH patients during autonomic testing. **Methods:** Fifteen controls (61±14 years) and 13 NOH patients (68±6 years;p=0.1) completed: 1)Deep Breathing (DB), 2) Valsalva maneuver (VM) and 3)Lower-body negative pressure (LBNP) during a functional MRI. Blood-oxygen level dependent (BOLD) contrasts were obtained and contrasted. **Results:** Compared to controls (C), patients (NOH) had significantly smaller heart rate (HR) responses to DB (C:15.3±9.6 vs.NOH:6.0±2.2) and VR's (C:2.1±0.47 vs.NOH:1.2±0.1;p<0.001). Patients had larger SBP drops during LBNP (C: -22.3±6 vs.NOH: -61±22) with significantly smaller compensatory tachycardias (19±8.5 vs.7.6±4.3)(p<0.001). BOLD response: During VM, controls had greater activation in the right (R) hippocampus (T-value:7.34), left (L) posterior cingulate (T-value:7.22) bilateral mid-cingulate (TR-value:5.76; TL-value:6.84) and bilateral thalamus (TR-value:7.23, TL-value:8.16) (pFWE<0.001). Following subtraction analysis, brain activation patterns showed no significant differences in the regions of interest in response to DB and LBNP. **Conclusions:** During tests of autonomic function, NOH patient had different cortical activation patterns during VM only. Cortical activation pattern during DB and LBNP showed similar patterns to that of controls.

P.067

Phosphoserine aminotransferase (PSAT) deficiency: Imaging findings in a child with congenital microcephaly

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Background: Serine deficiency disorders can result from deficiency in one of three enzymes. Deficiency of the second enzyme in the serine biosynthesis pathway, 3-phosphoserine aminotransferase (PSAT), has been reported in two siblings when the eldest was investigated for acquired microcephaly, progressive spasticity and intractable epilepsy. **Methods:** Our patient had neurological symptoms apparent at birth. Fetal magnetic resonance imaging (MRI) at 35 weeks gestation demonstrated microencephaly and simplification of the the gyration (anterior>posterior) which was confirmed upon subsequent post-natal MRI. Congenital microcephaly was apparent at birth. **Results:** PSAT deficiency was confirmed when exome sequencing identified biallelic mutations in *PSAT1*; c.44C>T, p.Ala15Val and; c.432delA, p.Pro144fs and biochemical testing noted low plasma serine 22 mcmol/L (normal 83-212 mcmol/L) and low CSF serine 10 mcmol/L (normal 22-61 mcmol/L). Despite oral serine and glycine supplementation at 4 months old the patient showed little neurodevelopmental progress and developed epileptic spasms at 10 months old. Serological testing for TORCH infections was negative. **Conclusions:** PSAT deficiency should be considered for patients with congenital microcephaly. Although further characterization of MRI findings in other patients is required, microencephaly with simplified gyral pattern could provide imaging clues for this rare metabolic disorder.

P.068

Hippocampal volume may predict early non-response to surgery in Trigeminal Neuralgia

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Background: Surgical treatment of trigeminal neuralgia (TN) can be highly effective, but durability of pain relief varies and factors influencing surgical failure are poorly understood. We hypothesized that structural brain differences—assessed using magnetic resonance

imaging (MRI)—might distinguish surgical responders from early non-responders. **Methods:** We retrospectively identified 35 TN patients treated surgically from 2005-2017 with high-resolution, pre-operative MRI scans adequate for quantitative structural analysis. Patients were classified as *non-responders* if, within 12-months after surgery, they: 1) underwent or were offered another surgical procedure; or 2) reported persistent, inadequately-controlled pain. Volumes of pain-relevant subcortical structures (amygdala, thalamus, and hippocampus) were measured on T1-weighted MRI scans using an automated approach (FSL-FIRST). **Results:** Surgical responders had significantly larger hippocampi bilaterally compared to early non-responders. Thalamus and amygdala volumes did not differ between groups. **Conclusions:** Pre-operative differences in brain structure, notably in the hippocampus, may predict durability of response to surgery in patients with TN.

Table 1: Demographic and Clinical Characteristics of TN Patients:

	Responders	**Non-Responders**	**P-value (2-tailed)**
Outcome Group	23	12	N/A
Sex (Female/Male)	9/14	6/6	0.5591
Age, years	4.35 ± 11.36	53.75 ± 16.33	0.9111
Affected Side (Left/Right)	5/18	5/7	0.2630
# of Previous treatments	0.13 ± 0.34	1.42 ± 1.40	0.0105*
Surgery Performed (MVD/PRR)	21/2	10/2	0.9722
Volumetric Assessment:			
	Responders (mm3)	**Non-Responders (mm3)**	**P-value (2-tailed)**
Hippocampus:			
Ipsilateral	3440 ± 365	697 ± 318	0.0415*
Contralateral	3381 ± 375	3727 ± 215	0.0015*
Left	3357 ± 373	3669 ± 231	0.0046*
Right	3464 ± 361	3754 ± 301	0.0178*
* p<0.05			
MVD – microvascular decompression surgery PRR – percutaneous retrogasserian rhizotomy			
Values are mean +/- standard deviation where appropriate			

Neuromuscular Disease and EMG

P.069

Respiratory dysfunction and sleep disordered breathing in children with Myasthenia Gravis

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Background: Myasthenia Gravis (MG) is an autoimmune disease that affects the neuromuscular junction. It typically presents with fluctuating muscle weakness which can affect respiratory muscles. Data about the prevalence of sleep disordered breathing in children with MG and the benefits of non-invasive ventilation outside the setting of MG crisis has not been studied so far. **Methods:** Eleven children between 3 and 18 years old with confirmed MG were recruited from the The Hospital for Sick Children Neuromuscular clinic in a prospective observational study. Informed consent was obtained and patients underwent PFTs, MIP/MEP, SNIP, FVC and standard polysomnography testing's. **Results:** In our study, we found that 2/11 children had abnormal Apnea Hypopnea index (AHI) and were diagnosed with obstructive sleep apnea (OSA). One of them has juvenile ocular MG with mild to moderate OSA and the second child has congenital MG with mild OSA. CPAP therapy was initiated for both patients. **Conclusions:** In our cohort, obstructive sleep apnea rate was significantly higher in children with MG than the known prevalence in general pediatric population (18% vs 2-3%). Early diagnosis and management of OSA can have great impact on children's health and quality of life. A larger study is needed to validate our findings.

P.070

Autosomal dominant MARS mutation linked to severe early onset CMT2U

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Background: Methionyl-tRNA synthetase (MARS) links methionine to its cognate tRNA required for translation. MARS mutations have been linked to adult-onset CMT2U. **Methods:** The proband had weakness in her first year of life, sitting at 11 months and walking at 20 months old. At 4 years old she was areflexic with distal > proximal weakness. Nerve conduction studies showed normal median and sural sensory responses with absent common peroneal, low median and tibial motor amplitudes. EMG noted denervation and quadriceps biopsy revealed neurogenic atrophy. Genetic testing for spinal muscular atrophy and sequencing of *MNF2, RAB7A, LMNA, MPZ, HSPB1, NEFL, GADP1, TRPV4, HSPB8, GJB1* and *PLEK8G5* were negative. She stopped walking at 9 years old and could not raise her arms above her head at 11 years old. **Results:** Exome sequencing identified *MARS*: c.1189G>A; p.Ala397Thr. To determine the functional consequences of p.A397T-*MARS*, yeast complementation assays were performed. Wild type or mutant *MARS* were cloned into yeast lacking the endogenous *MARS* ortholog. Wild-type *MARS*