morphic xanthoastrocytoma were high, but the Ki-67 labeling index was 1%. In the ganglioglioma, the T/N ratio of FLT was high, but the T/N ratio of MET was low. CONCLUSION: Specialized multiple PET accumulation patterns for tumors are useful for discriminating each tumor.

IMG-03. RESPONSE ASSESSMENT IN PEDIATRIC LOW-GRADE GLIOMA: RECOMMENDATIONS FROM THE RESPONSE ASSESSMENT IN PEDIATRIC NEURO-ONCOLOGY (RAPNO) WORKING GROUP

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INTRODUCTION: Pediatric low-grade gliomas (pLGG) show clinical and biological features that are distinct from their adult counterparts. Consequently, additional considerations are needed for response assessment in children compared to the established adult Response Assessment in Neuro-Oncology (RANO) criteria. Standardized response criteria in pediatric clinical trials are lacking, complicating comparisons of responses across studies. We therefore established an international committee of the Radiologic Assessment in Pediatric Neuro-Oncology (RAPNO) working group to develop consensus recommendations for response assessment in pLGG. METHODS: The committee consisted of 25 international experts in the areas of Pediatric Neuro-Oncology, Neuroradiology and Neurosurgery. The committee first developed a set of agreed upon topics they deemed necessary to understand the controversies of imaging utilization and assessment in pLGG. These topics were divided up among the committee members who presented all available literature to the entire RAPNO committee via web teleconference. Once presented, the group discussed these data and developed consensus statements and recommendations based on available literature, committee expertise and clinical experience. Each topic was discussed until a consensus was reached. RESULTS: Final consensus included recommendations about the following topics: specific imaging sequences, advanced imaging techniques, NF1-associated pLGG, molecular and histologic classification, assessment of cysts, vision and other functional outcomes as well as overall radiologic response assessment. CONCLUSIONS: The RAPNO pLGG consensus establishes systemic recommendations that represent an initial effort to uniformly collect and assess response in pLGG. These recommendations should now be evaluated internationally and prospectively in an effort to assess clinical utility, validate and modify as appropriate.

IMG-04. RESPONSE ASSESSMENT IN PEDIATRIC HIGH-GRADE GLIOMA: RECOMMENDATIONS FROM THE RESPONSE ASSESSMENT IN PEDIATRIC NEURO-ONCOLOGY WORKING GROUP

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INTRODUCTION: Response criteria for pediatric high-grade gliomas (pHGG) have varied historically and across clinical trials. Compared to adult HGG, pHGG response assessment has unique challenges. An international Response Assessment in Pediatric Neuro-Oncology (RAPNO) working group was established to develop pHGG response assessment criteria. METHODS: Pediatric and adult neuro-oncologists, neuro-radiologists and experts in imaging informatics developed a consensus statement and established a unified response assessment for biopsy-proven pHGG, excluding DIPG. This was achieved by identifying major challenges, reviewing existing literature and current practices, and finally developing recommendations through an iterative process. RESULTS: Categories for response assessment include complete response, partial response, minor response, stable disease and progressive disease. Refractory disease is excluded. Criteria used to determine response assessment include quantitative evaluation of measurable disease, qualitative assessment of diffusion imaging, presence or absence of new lesions, clinical status using performance score, and vascular endothelial growth factor inhibitor and/or corticosteroid use. Response is determined over 2-time points ≥ 8 weeks apart, and when progressive disease is unclear, guidance for repeat MRI imaging and/or utility of repeat biopsy is described. A number of recommendations are also given to standardize response assessment across clinical trials including MRI protocol sequence recommendations for brain and spine, definitions for measurable and non-measurable disease, and imaging time points with post-operative considerations. In addition, guidance is given for differentiating vasogenic edema versus tumor invasion in non-enhancing disease. CONCLUSION: Consensus recommendations and response definitions have been established and, similar to other RAPNO recommendations, prospective validation in clinical trials is warranted.

IMG-05. INITIAL RADIOGRAPHIC ASSESSMENT OF DWI AND ADC VALUES IN CHILDREN AND YOUNG ADULTS TREATED WITH DAY101 (TAK-580) FOR RECURRENT LOW-GRADE GLIOMAS (LGG) HARBORING MAPK ALTERATIONS

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BACKGROUND: Apparent diffusion coefficient (ADC) is a quantitative measure reflecting observed net movement of water calculated from a diffusion-weighted image (DWI), correlating with tumor cellularity. The higher cellularity of high-grade gliomas results in diffusion restriction and reduced ADC values, whereas the lower cellularity of low-grade gliomas (LGGs) gives higher ADC values. Here we examine changes in ADC values in patients with LGGs treated with the type 2 RAF inhibitor DAY101 (formerly TAK580). METHODS: Historical, baseline, and on-treatment brain MRIs for 9 patients enrolled on a phase 1 study of DAY101 in children and young adults with radiographically recurrent or progressive LGG harboring MAPK pathway alterations were obtained, de-identified and independently evaluated for ADC changes. Time points included baseline, first follow-up, and best response. Data processing of ADC estimates was performed using pmod molecular image software package. ADC changes were displayed as a histogram with mean values. Results were based upon a single read paradigm. RESULTS: There was a clear shift to lower ADC values for the solid component of tumors, reflecting changes in cellularity and tissue organization, while necrosis correlated with a shift toward higher ADC values. DWI

reveals reduced ADCs in responding tumors, with the percent change in ADC from baseline correlating with deeper RANO responses. CONCLUSION: DWI analysis reveals reductions in ADC values that correlates with treatment response and a shift toward more normal cellularity in tumors treated with DAY101. Changes in ADC may represent a novel imaging biomarker, reflecting biological response to DAY101 treatment.

IMG-06. PREDICTING SURVIVAL FROM PERFUSION AND DIFFUSION MRI BY MACHINE LEARNING

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INTRODUCTION: Magnetic Resonance Imaging (MRI) is routinely used in the assessment of children's brain tumours. Reduced diffusion and increased perfusion on MRI are commonly associated with higher grade but there is a lack of quantitative data linking these parameters to survival. Machine learning is increasingly being used to develop diagnostic tools but its use in survival analysis is rare. In this study we combine quantitative parameters from diffusion and perfusion MRI with machine learning to develop a model of survival for paediatric brain tumours. METHOD: 69 children from 4 centres (Birmingham, Liverpool, Nottingham, Newcastle) underwent MRI with diffusion and perfusion (dynamic susceptibility contrast) at diagnosis. Images were processed to form ADC, cerebral blood volume (CBV) and vessel leakage correction (K2) parameter maps. Parameter mean, standard deviation and heterogeneity measures (skewness and kurtosis) were calculated from tumour and whole brain and used in iterative Bayesian survival analysis. The features selected were used for k-means clustering and differences in survival between clusters assessed by Kaplan-Meier and Cox-regression. RESULTS: Bayesian analysis revealed the 5 top features determining survival to be tumour volume, ADC kurtosis, CBV mean, K2 mean and whole brain CBV mean. K-means clustering using these features showed two distinct clusters (high- and low-risk) which bore significantly different survival characteristics (Hazard Ratio = 5.6). DISCUSSION AND CONCLUSION: Diffusion and perfusion MRI can be used to aid the prediction of survival in children's brain tumours. Tumour perfusion played a particularly important role in predicting survival despite being less routinely measured than diffusion.

IMG-07. GADOLINIUM IS NOT NECESSARY FOR SURVEILLANCE MR IMAGING IN CHILDREN WITH CHIASMATIC-HYPOTHALAMIC LOW GRADE GLIOMA

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BACKGROUND: Patients with chiasmatic-hypothalamic low grade glioma (CHLGG) have frequent MRIs with gadolinium based contrast agents (GBCA) for disease monitoring. Cumulative gadolinium deposition in children is a potential concern. The purpose of this research is to establish whether MRI with GBCA is necessary for determining tumor progression in children with CHLGG. METHODS: Children with progressive CHLGG were identified from Texas Children's Cancer Center between 2005–2019. Pre- and post-contrast MRI sequences were separately reviewed by one neuroradiologist who was blinded to the clinical course. Three dimensional measurements and tumor characteristics were collected. Radiographic progression was defined as a 25% increase in size (product of two largest dimensions) compared to baseline or best response after initiation of therapy. RESULTS: A total of 28 patients with progressive CHLGG including 683 MRIs with GBCA (mean 24 MRIs/patient; range: 10–43 MRIs) were reviewed. No patients had a diagnosis of NF1. Progression was observed 92 times, 91 (98.9%) on noncontrast and 90 (97.8%) on contrast imaging. Sixty-seven radiographic and/or clinical progressions *necessitating management changes* were identified in all (100%) noncontrast sequences and 66 (98.5%) contrast sequences. Tumor growth >2 mm in any dimension was identified in 184/187(98.4%) on noncontrast and 181/187(96.8%) with contrast imaging. Non primary metastatic disease was seen in seven patients (25%), which were better visualized on contrast imaging in 4 (57%). CONCLUSION: MRI without GBCA effectively identifies patients with progressive disease. One should consider eliminating contrast in imaging of children with CHLGG with GBCA reserved for monitoring those with metastatic disease.

IMG-08. UNUSUAL IMAGING FINDINGS IN TWO CASES OF PAEDIATRIC LOW GRADE GLIOMA

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Low grade gliomas (LGG), including pilocytic astrocytoma (PCA), are the commonest paediatric brain tumours and their behaviour is well understood, typically following a benign course. BRAF fusion is common, particularly in PCA of the cerebellum and optic pathway. Here we present two patients whose LGG behaved in an unusual fashion. The first patient who was treated 6 years previously on LGG2 with vincristine and carboplatin for a tectal plate lesion was identified on routine imaging to have local tumour progression and underwent completion staging. This showed a new enhancing soft tissue abnormality within the spinal cord at the level of L2. Due to radiological dubiety both lesions were biopsied for histological and molecular analysis, confirming LGG of the tectal plate and finding the spinal lesion to be a myxopapillary ependymoma. The second patient presented with acute hydrocephalus following a 2 year history of neurocognitive impairments. He was found to have a large, complex tumour centred in and expanding the bodies of both lateral ventricles with significant mass effect. Radiologically this was most in keeping with a central neurocytoma but histological analysis confirmed it to be a PCA with KIAA1549-BRAF fusion. The first case demonstrates the utility of molecular analysis in confirming two distinct tumour types in one patient, in a situation where metastasis would not be expected and would significantly alter treatment and prognosis. The second is an example of how imaging can be misleading in a KIAA1549-BRAF fused PCA presenting as an intraventricular mass.

IMG-09. RESPONSE ASSESSMENT IN DIFFUSE INTRINSIC PONTINE GLIOMA (DIPG): RECOMMENDATIONS FROM THE RESPONSE ASSESSMENT IN PEDIATRIC NEURO-ONCOLOGY COMMITTEE

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Optimizing the conduct of clinical trials for diffuse intrinsic pontine glioma (DIPG) involves use of consistent, objective disease assessments and standardized response criteria. The Response Assessment in Pediatric Neuro-Oncology (RAPNO) committee, an international panel of pediatric and adult neuro-oncologists, clinicians, radiologists, radiation oncologists, and neurosurgeons, was established to address unique challenges in assessing response in children with CNS tumors. A subcommittee of RAPNO was formed to specifically address response assessment in children and young adults with DIPG and to develop a consensus on recommendations for response assessment. Distinct issues related to the response assessment of DIPG include its definition and recent molecular classifications, dearth of imaging response data, the phenomena of pseudoprogression, and measuring response in the era of focal drug delivery. The committee has recommended response be assessed using magnetic resonance imaging (MRI) of brain and spine, neurologic examination, and use of supportive medication, i.e. steroids and anti-angiogenic agents. Clinical imaging standards and imaging quality control