Within each TMZ cycle 5 days of immunogenic cell death (ICD) therapy (5 injections with Newcastle Disease Virus and 5 sessions of modulated electrohyperthermia (Oncotherm 50 min 40-60 Watt) was added at days 8 to 12. After all chemo-/ICD-therapy we continued with active specific immunotherapy: two autologous mature monocyte-derived dendritic cell vaccines loaded with ICD therapy-induced serum-derived antigenic extracellular microvesicles and apoptotic bodies (IO-Vac®). One month after the second IO-Vac®, 17 months after diagnosis, a temporal right FLAIR-visible region showed expansion, and three months later also diffuse contrast enhancement, which was confirmed in a control scan one month later. The original tumor was meanwhile reduced to 16 cm^3. However, in the last available scan, two months after the former, the contrast enhancement was disappeared, and the pathologic area on FLAIR was diminished. The original tumor size was reduced to 2 cm^3, two year after first diagnosis. She showed allergic skin reactions to TMZ, which was covered with systemic histamine intake. There were no side effects related to multimodal immunotherapy. Transient MRI changes can be observed even in distance from the original tumor and can be interpreted as immune-mediated effects when the original tumor is responding.

IMG-08. RESPONSE ASSESSMENT FOR PEDIATRIC CRANIOPHARYNGIOMA: RECOMMENDATIONS FROM THE RESPONSE ASSESSMENT IN PEDIATRIC NEURO-ONCOLOGY (RAPNO) WORKING GROUP

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INTRODUCTION: Craniopharyngioma (CP) is a histologically benign tumor of the pituitary stalk that accounts for 4% of pediatric central nervous system (CNS) tumors. Given its location, CP often causes neuro-endocrine, hypothalamic, and vision dysfunction. Standard therapy consists of maximally safe resection +/- radiation therapy (RT). Medical management, including intra-cystic therapy, may have utility in certain contexts. Survival after CP is excellent, but quality of life is often poor secondary to functional deficits from the tumor and/or treatment. Few prospective CP trials have been performed, and response assessment has not been standardized. METHODS: The Response Assessment in Pediatric Neuro-Oncology (RAPNO) committee, formed of international experts in relevant subspecialties, devised consensus guidelines from published literature and/or expert opinion to assess CP response in prospective clinical trials. RESULTS: Magnetic resonance imaging (MRI) is the recommended radiological modality for baseline and follow-up CP assessment. Computed tomography can be useful for identification of calcification in the initial diagnostic work-up. The committee defined specific standard MRI-based sequences focused on comprehensive evaluation of the suprasellar space. Radiologic CP response is defined by two-dimensional measurements of both solid and cystic tumor components. Three-dimensional measurements are also encouraged in prospective trials. In certain clinical contexts, response of solid and cystic disease may be differentially considered based on their unique natural histories and responses to treatment (including transient cyst growth during or after RT). Importantly, the committee incorporated functional endpoints related to neuro-endocrine and visual assessments into CP response definitions. In most circumstances, cystic disease should be considered progressive only if growth is associated with acute, new-onset or progressive functional impairment. CONCLUSION: CP is a common pediatric CNS tumor for which standardized response parameters have not been defined. A RAPNO committee devised guidelines for baseline and longitudinal assessments of CP to uniformly define response in future prospective trials.

IMG-09. CHARACTERISATION OF A PANEL OF *IN VIVO* MODELS OF PAEDIATRIC-TYPE DIFFUSE HIGH-GRADE GLIOMA (PDHGG) USING MAGNETIC RESONANCE IMAGING

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Novel therapies for paediatric-type diffuse high-grade glioma (PDHGG) are urgently required. Orthotopic models using patient-derived material are invaluable tools in preclinical drug development as they retain key genetic/epigenetic features, eg. histone H3G34 or H3K27 alterations. Their evaluation *in situ* is vital and requires sensitive imaging techniques such as MRI. 12 diffuse hemispheric glioma (DHG; 2 DHG-G34) and 21 diffuse midline glioma (DMG; 17 DMG-K27M) tumours have been characterised using MRI following site-specific orthotopic implantation of patient-derived cells directly from tumour material or after minimal expansion as stem cell cultures. Of the 62 models implanted; 3 DHG and 10 DMG samples were not tumourigenic and 13 DHG/3 DMG models are currently under MRI surveillance. Tumours identified on T_2-weighted (T_2w)-images varied from a diffuse hyperintense signal to well-defined high contrast masses. Tumour growth in 5 DMG models was too diffuse for longitudinal monitoring with T_2w-MRI. Once established, diffusion-weighted, T_1/T_2 mapping and contrast-enhanced MRI were used to further assess tumour phenotype. Quantitative data from 15 DMG models demonstrated higher water diffusivity and T_2 than 10 DHG tumours, which suggests less tightly packed tumour cells but may also reflect the closer proximity of tumours growing in the thalamus/pons/cerebellum to the ventricular system. Lack of contrast-agent enhancement in 11 DMG and 6/10 DHG models indicated an intact blood-brain barrier (BBB), with heterogeneous disruption observed in 4 DHGs; H3-G34 had no bearing on BBB integrity. Upon serial re-implantation survival was shortened in 3/4 DHG and 2/6 DMG models, while quantitative MRI parameters remained similar. Likewise, when 2 DHG and 2 DMG models grown in 2D/3D *in vitro* were implanted in parallel, poorer survival/improved penetrance was associated with 3D-cultured cells with no difference in imaging phenotype. The study highlights the potential of non-invasive MRI to accurately evaluate the efficacy of novel therapeutics in these PDHGG models.

IMG-10. DETERMINING BRAIN TUMOUR GRADE NON-INVASIVELY USING A SIMPLIFIED MRI PERFUSION PROTOCOL: SINGLE-BOLUS, LEAKAGE-CORRECTED DYNAMIC SUSCEPTIBILITY-CONTRAST MRI

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INTRODUCTION: Perfusion is associated with grade and survival in children's brain tumours. Dynamic susceptibility-contrast (DSC-) MRI measures perfusion non-invasively, estimating relative cerebral blood volume (rCBV). We previously showed significant differences between pre-treatment rCBV in low- and high-grade tumours in a multicentre study. Contrast agent leakage from tumour vessels during acquisition affects rCBV accuracy. A contrast agent pre-bolus can be given but this can be challenging in a clinical environment, introducing variability. Alternatively, a single bolus can be administered with leakage correction applied when processing the data. We investigated pre-treatment rCBV values in a multicentre study without pre-bolus administration. METHODS: Thirty-six patients underwent pre-treatment DSC-MRI scans at 2 centres on 4 different scanners. Protocols were variable. Pixel-by-pixel contrast agent concentration time courses were analysed. Maps of uncorrected ($rCBV_{uncorr}$) and leakage-corrected rCBV ($rCBV_{corr}$) were produced. Whole-tumour regions-of-interest were defined and median whole-tumour DSC-MRI parameters calculated. Patients subsequently underwent surgery / biopsy. Tumours were classified and graded. RESULTS: Twelve tumours were classified as low-grade; 24 as high-grade. Median whole-tumour $rCBV_{uncorr}$ was significantly higher in high-grade tumours than in low-grade tumours (1.628 *vs* -0.167, $p<0.001$). Median rCBV significantly increased in low-grade tumours following leakage correction (-0.167 to 1.072, $p=0.007$); there was no significant change for high-grade tumours. Using the median $rCBV_{uncorr}$ of 1.19 to differentiate between low- and high-grade tumours resulted in sensi-