Abstracts

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EPEN-29. IS THE OUTCOME OF SUPRATENTORIAL EPENDYOMA WITH ZFTA FUSION REALLY POOR?
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BACKGROUND: Survival of children with intracranial ependymoma is known to depend, in part, on the molecular group of the tumor. Previous studies have linked both PFA and ZFTA molecular group ependymomas to poor outcomes. As part of an unplanned analysis from ACNS0831, we explored outcomes for ZFTA supratentorial (ST) and PFA ependymomas.

METHODS: Patients were stratified by location, grade and extent of resection. Grade 2, ST tumors with no evidence of disease were observed only, N=37. All other patients were randomized to RT versus RT + maintenance chemotherapy or non-randomly assigned to RT + M if residual disease. A planned "as treated" analysis was performed by actual treatment received for randomized patients, excluding those who did not receive RT. Molecular groups were compared using two-sided log-rank tests stratified by study arms for all and randomized patients. Molecular characterization was achieved in 161/166 (97%) ST and 282/283 (99.6%) PE. Among ST, 94% were classified ZFTA and 6% YAP1. Homozygous CDKN2A deletion was detected in 16 tumors, all ZFTA and WHO grade 3. Among posterior fossa ependymomas, 88% were classified as PFA and 12% as PFN.

RESULTS: There was no statistical difference in outcome according to treatment. Across all study arms, 5-year EFS was 69.6%±4.3 for ZFTA vs. 52.9%±3.4 for PFA (p=0.028). For the randomized cohort, 5-year EFS was 72.1±4.8 for ZFTA vs. 60.1±3.9 for PFA (p=0.076). For the randomized cohort with initial GTRNTR, 5-year EFS was 74.1±5.1 for ZFTA patients vs. 61.0±4.2 for PFA patients (p=0.045). The 5-year EFS was 66.7±12.2 for ZFTA patients with CDKN2A deletion vs. 69.3±4.7 for ZFTA patients without CDKN2A deletion (p=0.232). CONCLUSION: Children with ST ZFTA ependymomas have a significantly better outcome than those with PFA tumors. Homozygous deletion of CDKN2A was present in 12% of ST ependymomas but was not an indicator of poor outcome.