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Molecular biological aspects of medulloblastomas in children under 3 years of age

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Medulloblastoma, embryonic neuroectodermal tumor of the cerebellum, is the most common malignant tumor of the central nervous system in children. Current therapy combines surgery resection, radiotherapy and chemotherapy. Significant advances in treatment were achieved by neurosurgical techniques improvement, radiation dose optimization and by introduction of the combined adjuvant chemotherapy. Treatment intensity depends on the child's age, extent of surgical resection and metastases presence. Current treatment schemes achieve a 5-year disease free survival in 70-81% of standard risk patients and in 30-70% of high-risk patients. Children under three years of age with medulloblastoma are primarily considered high-risk patients. The use of radiotherapy in the postoperative treatment is limited in this age group due to central nervous system immaturity. According to the new molecular biology findings, there are 4 "biological" medulloblastoma subgroups, known as WNT, SHH, group 3 and class 4. SHH tumors are found approximately in 60% of cases in this age group, group 3 tumors in 30% of cases. There is a strong correlation between SHH tumors and desmoplastic variant. Histological subtype (desmoplastic vs. classic variant) and molecular subgroup are the most important prognostic factors in children under 3 years of age. Based on a meta-analysis, the survival of children with desmoplastic medulloblastoma is significantly higher than with classic variant (about 80% vs. 25-40%). Postoperative treatment of children with medulloblastoma under three years of age therefore includes induction and consolidation chemotherapy (in the form of high-dose chemotherapy, mainly by thiotepa and carboplatin, supported by autologous haematopoetive drugs). Radiotherapy is not indicated in children with desmoplastic medulloblastoma, focal eventually craniospinal radiotherapy is considered individually in children with classic tumor subtype. We present a set of 7 patients under three years of age with medulloblastoma, treated at the University Hospital Brno in the last ten years. Desmoplastic subtype was the favorable prognostic sign in our group. The need of surgery performed in a comprehensive cancer center, with adequate tumorous tissue processing and biological testing should be emphasized, also the participation in international clinical trials with the possibilities of reference histopathological examinations and expert consultations. Also, more importance is currently given to biological treatment options.