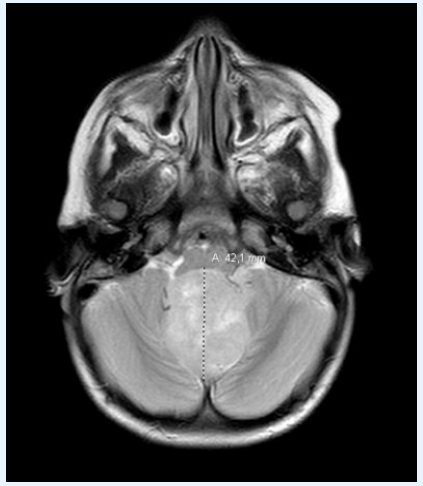
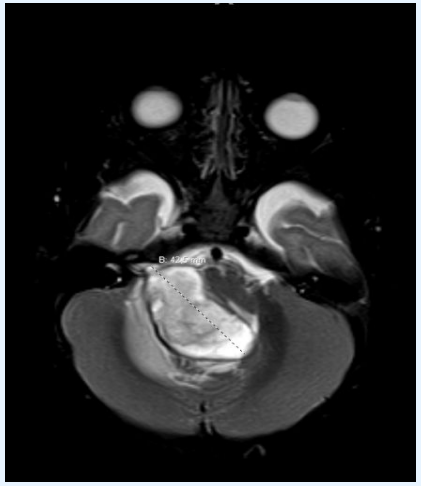
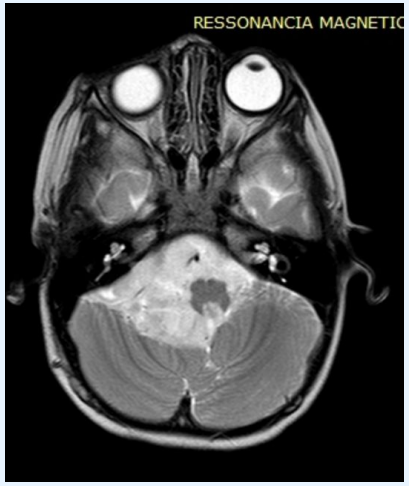
Survival analysis and prognostic factors in posterior fossa ependymomas in children and teenagers

Carolina Torres Soares, Marcos Devanir Silva da Costa, Patrícia Alessandra Dastoli, Jardel Mendonça Nicácio, Andrea Cappellano, Nasjla Sabá Silva e Sergio Cavalheiro.

Introduction and objective: Central Nervous System (CNS) tumors represent the most common type of solid tumor in the pediatric age group. Among these tumors, the ependymomas - the 3rd most common neoplasm for this site and age group stands out, and can be manifested mainly in three topographies: supratentorial, posterior fossa and spinal cord. More recent histopathological and molecular analyses define into subgroups according to the Classification of CNS tumors of the World Health Organization (WHO): subependymomas, myxopapilar ependymoma, supratentorial ependymoma, supratentorial - ZFTA fusion-positive, supratentorial - YAP1 fusion-positive, posterior fossa ependymoma, posterior fossa - group PFA, posterior fossa - group PFB, spinal ependymoma and spinal - MYCN-amplified. Currently, the most effective treatment is the maximum surgical resection followed by radiotherapy, however, ependymomas vary its behavior according to its location, its histological degree and the age of the patient at diagnosis. Thus, it seeks to analyze the overall and progression-free survival of patients under 18 years of age diagnosed with posterior fossa ependymomas, observing prognostic factors such as the degree of surgical resection, tumor topography, involvement of the lesion in the rhombencephalon disease, primary site of neoplasia, molecular classification of ependymoma and age group. **Methods:** For this retrospective cohort study, we accessed archives and medical records of patients under 18 years of age, treated at the institution after 2002 with diagnosis of ependymomas. Statistical analysis was performed by obtaining Kaplan-Meyer survival curves and Logrank Mentel-cox mathematical tests when two groups were compared or Logrank test for trend when more than two groups were compared. The qualitative variables were analyzed by contingency tests, using the Chi-square and Fisher's Exact tests, depending on the sample size and the adequacy of the normal distribution for each set of samples. It was considered α less than or equal to 0.05 for significance.







Results: According to the statistical analysis, the morphology of hindbrain involvement by the neoplasm implies in significant difference in progression-free survival – the median survival for patients with dorsal only involvement was 28 months, while for dorsolateral involvement it was 15 months and for total involvement, 9,5 months, p=0,0464 using Logrank test for trend. Despite that, overall survival for these groups showed no statistical difference – 44 months for dorsal involvement, 74 for the dorsolateral group and 31,5 for the total involvement group, using Logrank test for trend. Then, the subgroup analysis revealed significant contrast in the proportion of gross total resection (73.08% 19/26) in the group of only dorsal involvement of the rhombencephalon when compared (0% 0/6) of the total involvement group, p=0.0019 in fisher's exact test. There was also a statistical difference between the proportion of total resection (60% 9/15) in the dorsolateral involvement group when compared (0% 0/6) of the total involvement group, p=0.0186 in fisher's exact test. **Conclusion**: We confirmed previous findings, such as the impact of the extent of resection on overall and progression-free survival. Moreover, we concluded that the pattern of involvement of the brainstem by the tumor at diagnosis can bring important information regarding the prognosis of the patient in relation to disease-free survival and, likewise, the total involvement of the brainstem impairs total surgical resection of these tumors.