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LOW GRADE GLIOMAS IN CHILDHOOD—EXPERIENCE OF ONE INSTITUTION

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Purpose. To retrospectively analyze the clinical presentation, treatment and outcome of children with low grade gliomas.
Methods. Fifty seven patients with the diagnosis of low-grade gliomas, admitted to our institution between April 1995 and January 2002 were analyzed. The median age was 8 years (range 1 to 19 years), there were 29 males and 28 females. The median interval between symptoms and diagnoses was 6 months (range 1 month to 9 years). The primary tumor site was cerebellar n = 16 (28%) cerebral hemispheric n = 16 (28%) deep midline structures n = 11 (20%), intramedullary spinal cord tumors n = 8 (14%) and tumors of the visual pathway n = 6 (10%). Gross total resection (GTR) was attempted mainly for cerebellar and cerebral hemispheric tumors and biopsy or less aggressive resection for other tumor sites.
Results. The surgical procedure was gross total resection in 31(55%) patients, partial resection in 18(32%) patients and biopsy in 7(13%) patients. One patient with optic pathway glioma had neurofibromatosis. Six patients received adjuvant radiotherapy for partial resection and 4 patients received radiotherapy at relapse or disease progression. Ten patients received chemotherapy with carboplatin and vincristine, for tumors in deep midline structures (n = 7), intramedulllary spinal cord (n = 2) and relapsed cerebral hemispheric (n = 1). Seven out of 10 patients who received chemotherapy are alive without disease progression. Four patients were lost to follow up. The overall survival in 6 y 10 m is 84% and progression free survival is 82%.
Conclusion. Overall survival in these patients is comparable to other series. Gross total resection was associated with better disease control. For patients with low grade gliomas not amenable to surgery and young children, chemotherapy offers a good alternative of treatment. The multidisciplinary approach is fundamental.

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CSF LEVELS OF AFP AND B-HCG IN INFANTS: DEFINING A NORMAL RANGE

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Introduction and Objectives. Approximately 6% of germ cell tumors (GCT) arise within the CNS. These tumors encompass a wide histologic spectrum making biopsy desirable, however, open or stereotactic biopsy may be associated with unacceptable risk. In this situation, the measurement of AFP and B-HCG in serum and CSF may identify the presence of immature germ cell elements. To date, there are no published reference intervals for AFP or B-HCG in the CSF of infants. We therefore measured CSF AFP and B-HCG in infants aged <4 months who did not have malignancy to determine these reference intervals.
Methods. AFP (n = 84 infants) and B-HCG (n = 10 infants) in CSF were measured by a microparticle enzyme immunoassay on an Axsym analyser. Samples with >5000 rbc's/ml were excluded. Paired plasma samples were obtained from some patients.
Results. B-HCG was present in low concentrations in normal CSF. The limited data showed no variation in concentration with age. Using ages corrected for prematurity, AFP measurements (IU/ml) showed:

Age No.	Mean	CSF AFP No	Mean	Plasma AFP
0–15 days	20	67(2–362)	14	5640 (137–135070)
16–30 days	13	37(4–81)	11	1683(357–6450)
31–60 days	27	3(1-13)	13	273(67–1308)
61–110 days	26	1(1–4)	16	55(2–447)

Conclusion. CSF AFP levels are much lower than plasma levels, and by age six weeks in most infants, the levels are close to those found in adults. Corrected for prematurity, all results at age >2 months were <5 IU/ml. These results have implications for the identification of CNS tumors, particularly congenital CNS tumors in infants, containing immature germ cell elements.

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EXTRAGONADAL GERM CELL TUMORS: RESULTS OF AIEOP TCG 91/98 PROTOCOLS

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Aim of the Study. To analyse the outcome of patients with extragonadal (eg) GCT, treated with TCG 91/98 Protocols: 45 pts were enrolled: 34 pts were females, 11 males; median age 17 months (range 1–137 months). Site of the T: sacrococcyx (sc) in 37, mediastinum in 2, retroperitoneum in 2, salivary gland in 1, parotidis in 1, neck in 1, vulva in 1. Stage (St): St I:3 pts, St II:3, St III:28, St IV:11; AFP level was >10000 ng/ml in 19 pts (not measured in 5). Complete (compl) resection (res) at diagnosis was performed in 6 pts, partial (part) res in 9 pts, biopsy (bio) in 30; 17 pts underwent to delayed surgery (part in 9, compl in 8) of the residue, whose istology was malignant in four. All pts were treated with 2–4 courses of Chemotherapy consisting of Carboplatin (400 mg/m^2/d, D1,2) Etopo side (150 mg/m^2/d, D1,2) Ifosfamide (1,5 g/m^2/d, D21,21), Vincristine (1,5 mg/m^2/d,D21) (not included in TCG 98) DActinomycin (1,5 mg/m^2/d, D 21).
Results. 3 pts died for sepsis, 2 were no responders, 16 had relapse: following relapse treatments, 6 are in II-Rem, 1 is alive with disease. Exitus for progression occurred in 11 pts (2 no responders, 9 relapsed): in these 11 pts, the site of the T was sc in 8, mediastinum in 2, parotidis in 1; 6 pts were in St III, 5 St IV; AFP was >10000 ng/ml in 5, surgery at diagnosis consisted of part. res. of the T in 4, bio in 7; delayed part. res. of malignant residue was performed in 4.
Conclusions. in TCG 91/98 Protocols, sc and mediastinal T, partially resected, with malignant residue, frequently presented a poor outcome: it is reasonable to suggest the early recognition of resistent cases followed by different chemotherapic regimen, with the aim of obtain a complete resection of the T, necessary for cure.

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CNS GERM CELL TUMORS (GCT) TREATMENT RESULTS FROM ONE INSTITUTION

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Aim. To assess treatment results of patients with GCT treated according to SIOP protocol and compare it with historical group.
Materials & Methods. 55 pts. with were treated. The historical group consisted of 29 pts treated 1987–96. Diagnosis was based on pathology of resected tumors. Germinoma was diagnosed in 22 pts and secreting germ cell tumor (SGCT) in 7. Al were irradiated to the craniospinal axis. Patients with SGCT received adjuvant chemotherapy (3 × PVB). Since 08.96 SIOP protocol was implemented. 26 pts were treated—11 pts with SGCT and 15 germinomas. In the SGCT group 5 pts underwen delayed surgery, 6 pts had CR after chemotherapy. In the non secreting group 2 pts underwent primary tumor resection 13 pts had biopsy, in all CR was observed after chemotherapy. In case of relapse second line chemotherapy was implemented.
Results. In the historical group 23 out of 29 pts are alive, median follow up 6 3/12. Si patients died (2 pts-SGCT, 4-germinomas), from disease recurrence. In the second group 24 out of 26 pts are alive. In the SGCT 10 out of 11 pts are alive, 9 in first C from 11/12 to 64/12, 1 in second CR 12/12 from relapse. One patient died of relaps 1/12 from completing radiotherapy. In the germinomas 13 pts are alive in first CR 4/12-65/12, one pt in second CR 28/12 from relapse. One patient died of reasons n related to disease. 5 yrs EFS for historical group is 79% and 66% in the current, OS 79% and 89% respectively.
Conclusions. No statistically significant differences were found in the results of treatment in both groups. Omitting craniospinal radiotherapy didn't jeopardize outcome. Second line chemotherapy is effective in obtaining remissions in relapse patients.