



آغا خان یونیورسٹی  
THE AGA KHAN UNIVERSITY

# Accepted Abstracts of First Pakistan Pediatric Neuro Oncology Symposium

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## Free Paper Abstracts

### Abstract 1

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**Abstract Title:** “Braf V600e Inhibitors for pediatric low grade and high-grade glioma: J.P Garrahan hospital experience.”

**Abstract Theme:** Pediatric Neuro Oncology

**Introduction:** The BRAF V600E point mutation plays a key role in the tumorigenesis of many low- and high-grade gliomas. Its inhibition is part of the innovative and promising therapies emerging in recent years. Knowing its efficacy and tolerance is essential in the molecular era.

**Objectives:** To analyze the role of BRAF V600E inhibitors in the paediatric population with gliomas harboring the BRAF V600E mutation.

**Material and methods:** Descriptive, single-center study; collecting clinical, imaging and molecular information from patients under treatment with BRAFV600E inhibitors in the Neuro-Oncology service of Hospital Garrahan.

**Results and conclusion:** 13 patients were treated with BRAF inhibitions (Vemurafenib=10, Dabrafenib=1, both=2) in the last 7 years: 9 were low grade gliomas (3 disseminated) and 4 high grade gliomas (1 disseminated). The median age at diagnosis was 8.63 years (0.89-14.04), and at start of target therapy was 11.62 years (3.64-15.42). All patients had a previous surgical procedure, and 92% had received another therapy prior BRAF inhibition: 83% conventional chemotherapy (in one case, up to 4 different protocols) and 33% radiotherapy. Under target therapy, tumour response was obtained in 76% of patients (with size reduction greater than 25%), and the best response was observed in the first 6 months of treatment in 70%. Four patients progressed under treatment (all high-grade gliomas) and 2 progressed shortly after stopping the inhibitor (both low grade gliomas). Tolerance was acceptable: 38% had grade 3-4 toxicity, with complete recovery after dose reduction or treatment suspension. With this study, the safety and efficacy of BRAFV600E inhibitors treatment in a paediatric population with gliomas is demonstrated.

**Key words:** BRAF, V600E, HGG, LGG, CNS tumor"

## Abstract 2

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**Abstract Title:** “Bridging the treatment gap in infant medulloblastoma: molecularly informed outcomes of a globally feasible regimen.”

**Abstract Theme:** Pediatric Neuro Oncology

**Background:** Infant medulloblastoma represents an enormous challenge in neuro-oncology, due to their simultaneous high-risk of recurrence and high risk of severe neurodevelopmental sequelae with craniospinal irradiation. Currently infant medulloblastoma are treated with intensified protocols, either comprising intraventricular methotrexate or autologous transplant, both of which carry significant morbidity and are not feasible in the majority of the world. We sought to evaluate the molecular predictors of outcome in a cohort of infants homogeneously treated with induction chemotherapy, focal radiation and maintenance chemotherapy.

**Methods:** In a retrospective analysis, 29 young children treated with a craniospinal irradiation sparing strategy from Hospital Garrahan in Buenos Aires were profiled using Illumina Human Methylation EPIC arrays and correlated with survival.

**Results:** Twenty-nine children (range, 0.3–4.6 y) were identified, comprising 17 sonic hedgehog (SHH), 10 Group 3/4, and 2 non-medulloblastomas. Progression-free survival (PFS) across the entire cohort was 0.704 (95% CI: 0.551–0.899). Analysis by t-distributed stochastic neighbor embedding revealed 3 predominant groups, SHH $\beta$ , SHH $\gamma$ , and Group 3. Survival by subtype was highly prognostic with SHH $\gamma$  having an excellent 5-year PFS of 100% (95% CI: 0.633–1) and SHH $\beta$  having a PFS of 0.56 (95% CI: 0.42–1). Group 3 had a PFS of 0.50 (95% CI: 0.25–1). Assessment of neurocognitive outcome was performed in 11 patients; the majority of survivors fell within the low average to mild intellectual disability, with a median IQ of 73.5.

**Conclusions:** We report a globally feasible and effective strategy avoiding craniospinal radiation in the treatment of infant medulloblastoma, including a robust molecular correlation along with neurocognitive outcomes.

**Keyword:** brain tumor, infant, medulloblastoma, SHH, radiation.

## Abstract 3

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**Abstract Title:** “The prevalence of central nervous system tumors at Chris Hani Baragwanath Academic Hospital, Paediatric Oncology unit between 2006 and 2015.”

**Abstract Theme:** Paediatric Neuro-Oncology

**Introduction:** Tumours of the central nervous system tumours (CNS) are a diverse group of disease that constitute the most common solid malignancy in children accounting for more than 20% of all childhood cancers in high-income countries. According to the SA paediatric cancer registry over a period of 25 years, CNS tumours in children were the 3rd commonest tumours following leukaemia and lymphoma.

**Objective:** To describe the demographics, spectrum and outcome of CNS tumours in children aged 0-19 years at a Paediatric Oncology Unit at a tertiary academic hospital in Johannesburg over a 10-year period, 2006-2015.

**Methods:** This was a health-facility based retrospective descriptive study. Children aged 0-19 years with primary CNS tumours seen at the POU during the study period were eligible. Metastatic CNS disease were excluded.

Data was analysed using Stata. The Kaplan- Meier survival estimates were used for 5-year survival rates. The outcome event was death. Patients without events were censored at their last follow up visit. Univariate Cox proportional hazards models were used to identify possible risk factors for death.

**Results:** CNS tumours were diagnosed in 169/1231 new patients during the 10-year period thus constituting 13.73% of all tumours. One hundred thirty-seven records were available for analysis. Mean age was 7.83 years s.d 0.4. The commonest tumours were astrocytic (44.5%), followed by craniopharyngioma (19.7%), medulloblastoma (14.6%), PNET (5.1%), pineoblastoma (3.6%) and others. The survival rates at 1 and 5 years were 69% and 51% respectively. Patients diagnosed with medulloblastoma or craniopharyngioma were less likely to die (p- value 0.008 and 0.007) whereas those with high grade tumours were 2.44 times more likely to die (p-value =0.013)

**Conclusion:** The spectrum of paediatric CNS tumours is similar to what has been reported in other HIC and LMIC with a high lost to follow up

**Recommendation:** Strengthen follow up measures.

## **Abstract 4**

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**Abstract Title:** “Incidence of Posterior Fossa Syndrome in children at a tertiary care hospital in Karachi.”

**Abstract Theme:** Pediatric Neuro Oncology

**Other Theme:** Posterior Fossa Syndrome

**Objective:** Pediatric brain tumors accounts for 25% of all cancers, out of which 60% arise in the posterior fossa. The most debilitating complication of posterior fossa surgery is posterior fossa syndrome (PFS) or cerebellar mutism syndrome. The overall incidence of PFS has been noted to be 11-29%. It is characterized by long-term neurological, speech-language, and cognitive impairment. The incidence of this serious neurosurgical complication remains incompletely defined in developing countries. This study aims to determine the incidence of PFS in children to reduce this post-surgical complication.

**Methods/Materials:** All children (<1year-18years) with a diagnosis of posterior fossa tumor, who had undergone surgical resection at Aga Khan University Hospital between January 2014 – June 2019 were reviewed retrospectively for demographics, symptoms, radiological findings, tumor characteristics and treatment of posterior fossa syndrome.

**Results:** 79 patients were included in the study. PFS developed in 56(70.9%) patients. Medulloblastoma 30(38%) was noted to be the most common tumor type associated with PFS. Most common tumor location was the 4th ventricle in 30(38%) patients and gross total resection was done in majority of the cases, 32(40.5%). Most common post-operative symptoms were ataxia 41(52%) followed by irritability 38(48%) and slurred speech 32(40%). Steroids were given to all patients pre and post operatively. 38(48%) patients were helped with physiotherapy and 18(23%) received zolpidem for improvement of symptoms.

**Conclusion:** Majority of patients (70.9%) experienced PFS after having posterior fossa surgical resection as compared to the data from the published literature. Collaborative team effort is highly warranted to prevent this serious post-surgical complication in children.

**Key words:** cerebellar mutism syndrome, Posterior fossa syndrome, CNS tumors"

## Abstract 5

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**Abstract Title:** “Defining the clinical and prognostic landscape of Embryonal Tumors with multi-layered rosettes (ETMRs), A Rare Brain Tumor Consortium (RBTC) Registry study.”

**Abstract Theme:** Pediatric Neuro Oncology

**Introduction:** "ETMRs, are rare, aggressive embryonal brain tumours. Encompassing various histological entities ETMRs were only recently recognized by WHO as distinct molecular entity. Characterized by C19MC-alterations ETMRs remain poorly understood as ~200 published reports lack molecular homogeneity. This has led to failure of disease pattern recognition and treatment modalities associated with best survival outcome.

We, therefore, undertook comprehensive molecular analysis of 623 CNS-PNETs enrolled in the RBTC with the goal of determining the clinical landscape of ETMRs.

We identified 159 ETMRs using combination of molecular analyses including, FISH (125), methylation (88), SNP and RNAseq (32); 91% had C19MC amplification/gains/fusions 9% lacked C19MC alterations but had global methylation features of ETMR-NOS.

ETMR, is a disease of infancy and present at median age of 26months. Unique to EMTRs is female preponderance (1:1.4). ETMRs arise in all CNS locations, notably 10% were brainstem primaries mimicking DIPG. Majority are localized (M0:72%), ETMRs can behave like soft tissue sarcoma's presenting with advanced metastasis (M2-3:18%).

Uni-and multivariate analyses of clinical and treatment details of curative regimens available for 110 patients identify metastasis ( $p=0.002$ ), brainstem locations( $p=0.005$ ), extent of resection, receipt of multi-modal therapy (high dose chemotherapy and radiation) ( $P<0.001$ ) as significant treatment prognosticators, while C19MC status, age and gender were non-significant risk factors. EFS at 3 (84%, 95%CI:77-91), 12 months (37%, 95%CI:20-41) and OS at 4yrs (27%, 95%CI:18-37) indicate despite intensified therapies ETMR is a rapidly progressive, fatal disease.

Our comprehensive analysis on the largest cohort of molecularly-confirmed ETMRs provides critical framework for improved diagnostic recognition, clinical management and prospective trial development."



## Abstract 6

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**Abstract Title:** “Risk Stratification of Medulloblastoma on the Basis of Molecular Subgrouping: An Experience from a Single Tertiary Health Care Centre from Developing Country.”

**Abstract Theme:** Pediatric Neuro Oncology

**Key words:** Medulloblastoma, molecular subgrouping, twinning

**Objective:** Medulloblastoma is the most common malignant childhood brain tumor and is a major cause of morbidity and mortality. Classically, medulloblastoma has been risk stratified on the basis of clinical characteristics (age, metastasis and extent of resection) and histological types (classic, desmoplastic and anaplastic). However, recently medulloblastoma has been subgrouped by using a variety of different genomic techniques (such as gene expression profiling, microRNA profiling and methylation array) into 4 groups (WNT, SHH, Group 3 and group 4). This new stratification has potentially important therapeutic and prognostic implications. There are very scarce data on the molecular sub grouping of medulloblastoma in developing countries. This is the only data from Pakistan on the clinical profile and molecular subgrouping in children with medulloblastoma.

**Methods/Materials:** All children (0 – 16 years) diagnosed with Medulloblastoma between April 2014 and January 2020 at Aga Khan University Hospital were reviewed retrospectively for clinical data, histology, molecular sub grouping, risk stratification, management and outcome. Biopsy samples of patients diagnosed to have medulloblastoma were sent for molecular sub grouping to Hospital for Sick Kids, Toronto as a part of a twinning program with Aga Khan University Hospital.

**Results:** Thirty-six children were included in the study. There were 28 (77.7%) males. Most of the patients were 3-9.9 years’ age group (52.7%). Most common presenting complains were vomiting & headache 33 (91.6%). Three patients (8%) had a family history of cancer. Metastatic disease was present in 10 (27.7 %) patients at diagnosis. Molecular subgrouping was done for 30 patients and showed Group 4 in 14, SHH in 7 (2 were p53 mutated), WNT in 4 patients. There were 3 patients with Group 3 and the subgrouping was inconclusive in 2 patients. Revised risk stratification based on extent of resection, metastasis and molecular subgrouping (Ramaswamy, Acta Neuropathol 2016) showed that there were 3 low risk patients with WNT subtype. There were 18 patients who were standard risk, 13 high risks and 2 very high risk. Thirty patients were treated with concomitant chemotherapy and radiation followed by maintenance chemotherapy. Twenty-nine completed therapy. One patient had progressive disease on therapy. There were 17 (47.2%) patients who had post-surgery cerebellar mutism, eleven patients expired, of which 2 expired during treatment and 4 had relapse after completion of therapy. Five patients expired without getting any treatment. Twenty-four patients are on regular follow up while one patient was lost to follow up.

**Conclusion:** These are the only data from Pakistan showing risk stratification based on molecular subgrouping. This had an important impact on the increase in the number of average risk patients and their consequent management and prognosis. Twinning programs can significantly contribute towards improvement in proper diagnosis and management of pediatric brain tumors.

## Abstract 7

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**Abstract Title:** “Pediatric intracranial Germ cell tumors, KHCC experience.”

**Abstract Theme:** Pediatric Neuro Oncology

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**Background:** Intracranial Germ Cell Tumors (iGCT) are rare and divided into germinomas and non-germinomatous germ cell tumors (NGGCT). Despite a good prognosis, the neurocognitive, visual and neuroendocrine long-term morbidities remain of concern.

**Methods:** We retrospectively reviewed children diagnosed < 18 years old with iGCT treated at KHCC /Jordan over 18 years. Clinical characteristics, management and morbidities are reported.

**Results:** Twenty-one children (15 males) with a median age 11.9 years (range,6.1-17.3 years) were identified. Thirteen patients had polyuria/polydipsia on presentation, visual complaints (9), headaches (7), and five had weight changes. Median duration of symptoms was 6 months (range,0.5-48 months).

There were 16 germinomas (13 males) and 5 NGGCT (2 males). Tumor location was suprasellar (11, males 7), pineal (7, males 6), bifocal (2 males) and quadrigeminal plate (1 female). Seven tumors (33%) were metastatic (7 ependymal, 3 also parenchymal). Twelve patients had elevated tumor markers and 15 patients underwent tumor biopsy.

Twenty children received chemotherapy; for germinoma 13 received carboplatin/etoposide and 2 received cisplatin/etoposide, while children with NGGCT all received carboplatin/etoposide/ifosfamide. All patients received radiotherapy, in germinoma (WVF 24Gy (6), whole brain 24Gy (4), WVF 24Gy with boost (3), CSI 24Gy (3)), while all NGGCT received 30-36Gy CSI with boost. All patients with germinoma are alive in remission except one lost to follow-up after relapse. Three of five children with NGGCT died (relapse (1), sepsis (1), electrolyte imbalance (1)).

With median follow-up of 6.9 years (range, 0.1-17.3 years) for 16 survivors; there was no change in the hormonal requirements relative to presentation (4 not on supplements), and vision was stable or better (1/28 eyes blind, 5 poor, 22 good). Four patients attended college (2 with mental challenges), three had average school performance and the rest had poor performance /left school.

**Conclusions:** Germinomas have excellent survival so it is important to limit long-term morbidities. Early recognition of symptoms and proper diagnosis without a surgical intervention, when possible, may minimize these morbidities.

**Key words:** intracranial germ cell tumor, developing country, quality of life.

## Abstract 8

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**Abstract Title:** “Combination of arginine depletion and polyamine inhibition as an anticancer strategy for diffuse intrinsic pontine glioma (DIPG).”

**Abstract Theme:** Pediatric Neuro Oncology

DIPG is an aggressive pediatric brainstem tumor, with a median survival below 12 months. Tumor cells are dependent upon arginine, an essential amino acid, metabolised by arginase enzymes into ornithine, a pivotal precursor to the polyamine pathway. Polyamines, frequently upregulated in cancer, are intracellular polycations controlling key biological processes – the inhibition of which we have previously shown to be highly efficacious in preclinical DIPG models. Pegylated arginase (BCT-100) has recently been shown to significantly delay tumor development, prolonging survival of neuroblastoma-prone Th-MYCN mice. This study investigated the effects of arginine depletion therapy as a single agent and in combination with polyamine pathway inhibitors in DIPG. We found that ARG2, the protein encoding for arginase, is expressed significantly higher in DIPG tumors compared to normal brain. Arginine depletion via BCT-100 reduced DIPG cell proliferation and colony formation in patient-derived cell lines. Using orthotopic patient-derived xenograft models of DIPG our results showed that frequent dosing of BCT-100 significantly delayed tumor development and increased the survival of the mice ( $p < 0.0001$ ). DFMO is an FDA-approved inhibitor of the enzyme ornithine decarboxylase, a key driver of polyamine synthesis. The combination of BCT-100 with DFMO led to a significant enhancement in DIPG survival ( $p < 0.005$  compared to single agent treatments). Triple combination therapy with the addition of the polyamine transport inhibitor AMXT-1501 led to a potent and profound improvement in survival. These data show that arginine depletion therapy using BCT-100 combined with dual polyamine inhibitory agents represents a potentially exciting new approach for the treatment of DIPG.

## Abstract 9

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**Abstract Title:** “Relapse pattern and quality of life in patients with localized basal ganglia germinoma receiving focal radiotherapy, whole-brain radiotherapy, or craniospinal irradiation.”

**Background and Purpose:** The optimal radiation field in localized basal ganglia (BG) germinoma is still undetermined. Thus, based on the relapse pattern and health-related quality of life (HRQOL), we evaluated three popular radiation fields. **Material and Methods:** The clinical data of 161 patients with localized BG germinoma were included in this retrospective study. Relapse status and relapse sites after treatment were explored. HRQOL was evaluated using the Pediatric Quality of Life Inventory 4.0 (PedsQL 4.0) ( $\leq 15$  years) and Short Form-36 (SF-36) ( $> 15$  years) questionnaires based on the patients’ age at last follow-up.

**Results:** After a median follow-up duration of 83 months (range, 20–214 months), 19 patients experienced relapse, including 15, 4, and 0 patients in the focal radiotherapy (FR) ( $n=35$ ), whole-brain radiotherapy (WBRT) plus boost ( $n=109$ ), and craniospinal irradiation (CSI) plus boost ( $n=17$ ) groups, respectively. The 5-year disease-free survival rates were 74.3%, 97.2%, and 100%, respectively ( $p < 0.001$ ). Among the 15 patients who relapsed after FR, 14 had positive radiological findings, including seven (50.0%) with lesions in the periventricular area and seven (50.0%) with frontal lobe lesions. Relapse in both these areas were significantly reduced by WBRT or CSI. HRQOL data were available for 69 patients, who generally scored low. Among 38 patients evaluated by SF-36, those receiving CSI had significantly lower mental component scores than those receiving WBRT ( $p=0.027$ ) or FR ( $p=0.011$ ).

**Conclusions:** Considering both disease control and HRQOL, WBRT is the optimal radiation field in our series. The relapse pattern identified in patients receiving FR is informative for further treatment volume optimization.

## Low Middle Income Country (LMIC) Oral Abstracts

### Abstract 10

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**Abstract Title:** “To assess the impact of dedicated Paediatric Neuro Oncological services in a developing country: a single institution, Pakistani experience.”

**Abstract Theme:** Pediatric Neuro Oncology

**Other Theme:** Dedicated Paeditric Neuro oncology Services

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**Background:** To see the impact of dedicated pediatric neuro-oncological services (DPNOS) in a resource limited setting. The outcome measures include numbers and pattern of referral in two distinct periods 1998-2013(15 years Era1) and 2014-2019 (6 years, Era2) when the DPNOS started. It is an established fact that dedicated, multidisciplinary services in pediatric neuro-oncology help in increase patient referral, contributes to better management and outcomes, reduced abandonment, better palliative and end of life care and improved patient and caregiver satisfaction.

**Material and Methods:** A retrospective chart review of patients from 2014-2019 (Era2, 6years) and their outcome compared with previously analyzed data from 1998-2013 (Era1, 15 years) when there was no DPNOS. IRB was approved and results were analyzed using SPSS.

**Results:** In Era1, 231 patients came to our facility compared to 270 in Era2, a drastic increase in referral. In Era1 total abandonment rate was 56% which was significantly reduced in Era2 (24%). In Era1 average time from symptoms till reaching to us was 6 months which reduced to 2 months in Era2. In Era1 most common tumors were Gliomas (31%) Medulloblastoma (21%) and ependymomas (13%). In Era2 Medulloblastoma (29%) Gliomas (20%) and ependymomas (13%). In Era1 CNS tumors were 7 % 231/3315) of all referral versus 6 % in Era2 (270/4715). In Era1, only 23% were offered treatment with curative intent with an overall survival of 10% whereas in Era2 it was 35% and 24%, respectively.

**Conclusion:** Implementation of a DPNOS had a major impact towards better care. It led to an increase referral, reduced abandonment, better overall survival and avoiding delay toward the diagnosis and treatment. A large number of children need effective palliative care from the beginning due to advanced, inoperable, metastatic disease at presentation."

## **Abstract 11**

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**Abstract Title:** “Pediatric Neuro-oncology Twinning Program: a collaboration between Pakistan and Canada over a six-year period.”

**Abstract Theme:** Pediatric Neuro Oncology

"Low- and middle-income countries sustain the majority of pediatric cancer burden, with significantly poorer survival rates compared to high-income countries. Central nervous system cancers are among the leading causes of pediatric cancer related mortality. Higher mortality rates in low- and middle-income countries can be attributed to lack of funds and expertise in all but a few hospitals. Collaboration between institutions in low- and middle-income countries and high-income countries is one of the ways to improve cancer outcomes. We describe our experience of a pediatric neuro-oncology twinning program which began in 2014, between the Hospital for Sick Children in Toronto, Canada and several hospitals in Karachi, Pakistan including the Aga Khan University Hospital, Indus Children Cancer Hospital, Shaukat Khanum Memorial Cancer Hospital and Combined Military Hospital Rawalpindi. A total of 406 patients were included in this program. We studied the epidemiology of tumors and treatment plans, both medical and surgical, for patients that were included in the twinning program. Early referrals, formation of a multidisciplinary team, changes in radiation doses, necessity of immediate post-operative scans and second look surgery were some of the changes due to this twinning. We observed a steadily decreasing trend in changes in management plans based on recommendations from the Hospital for Sick Children, decreasing from 56% in 2014 to 12% in 2020. The average percentage of plan changes over the last six years was 22%. This video-teleconferencing twinning program has improved management of children with brain tumors. We believe multidisciplinary and collaborative efforts by experts from across the world have aided in the diagnosis and treatment of patients and helped establish local treatment protocols.

**Keywords:** twinning, pediatrics, neuro-oncology"

## Abstract 12

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**Abstract Title:** “A Retrospective Study of Pediatric Brain Tumors from a Tertiary Care in South China.”

**Objective:** To analyze the diagnosis, treatment, multidisciplinary team (MDT) work of childhood brain tumor treated at a tertiary children's hospital in South China and to better understand the challenges and opportunities in resource-limited district. **Methods:** A hundred and seventy-six cases were included at Shenzhen Children's Hospital (SZCH) from January 2015 to December 2018. The distributions of patients, pathology, comprehensive care, causes of death were retrospectively studied. **Results:** The numbers of patients were increased steadily from 2015 to 2018 which were 31, 38, 54 and 53, respectively. The male to female was 1.65 to 1. The median age was 6.6 years. In sum, 152 cases (86%) received operations and the others were diagnosed based on radiological findings and clinical manifestations due to difficulties in obtaining pathological diagnoses. The most common pathological types were glioma, medulloblastoma and ependymoma, accounting for 27%, 20% and 10% of patients, respectively. The treatment for brain tumor included operation, radiation, chemotherapy and autologous hematopoietic stem cell transplantation. A total of 75 patients required irradiations, accounting for 43% (75/176) of patients. Patients received irradiation outside SZCH, hence 28 patients lost long-term follow-ups. Three patients diagnosed with pinealoma, AT/RT and CNS embryonal tumor undergone autologous peripheral blood stem cell transplantation. Two melanoma patients received targeted therapy and one of them remained in stable condition. Thirty-four patients died due to disease progression, relapse, or severe complications. **Conclusion:** There is a trend increase of pediatric neuro-oncological patients in recent years. Options of adjuvant treatment have been implemented started from stereotactic and conformal radiation, to chemotherapy, to high dose chemotherapy with stem cell rescue, and to targeted treatment. However, long-term outcomes of these patients need further investigation.

## Abstract 13

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**Abstract Title:** “Pediatric Brain Tumors – Experience from a Tertiary Care Hospital.”

**Abstract Theme:** Pediatric Neuro Oncology

**Other Theme:** Clinicopathological profile and outcome of Pediatric CNS tumors

**"Background:** Central nervous system (CNS) tumors are the most common solid tumor in children under the age of 15 years. Pediatric neuro oncology is still at a very nascent stage in the developing countries and morbidity associated with CNS tumors may be significant in terms of physical deficits as well as neuropsychological and neuroendocrine sequelae. There are only a few reports on the multidisciplinary approach and outcomes of pediatric brain tumors in developing countries.

**Aims:** This study aims to identify the clinicopathological profile and outcome of Pediatric CNS tumors in a tertiary care center

**Materials and Methods:** A retrospective study of clinical, radiological & pathological profile, and outcomes of children <18 years diagnosed with brain tumors at our institute from October 2018 to October 2020 was done. Histopathological categorization was done as per the WHO classification 2007. Statistical Analysis was done by SPSS version 22.

**Results:** Total 118 patients were diagnosed with 55% male. Highest incidence was in the age group of 5-10 years. The most common diagnosis was medulloblastoma (27%) followed by pilocytic astrocytoma (19%), craniopharyngioma (10%), ependymoma (10%), GBM (10%), brain stem glioma (10%), other high grade gliomas (5%), other low grade gliomas (4%), multiple brain SOL (1.25%), choroid plexus tumors (1.25%), meningioma (1%), and CNS Lymphoma (2%). Amongst them, 50% received treatment (resection, radiotherapy and chemotherapy), 10% are on current waiting list, 4% were offered palliation due to inoperable tumors, and 36% abandoned/died. Regarding overall outcome, 31% are on follow up/active treatment, 37% lost to follow up, while 32% died.

**Conclusions:** brain tumors are still the leading cause of cancer related morbidity and mortality in children. For brain tumor management multidisciplinary team is essential. Higher rates of treatment abandonment are documented in view of complexity of the treatment with long duration, involving neurosurgery, radiation, chemotherapy, and high cost of treatment.



## Moderated Poster Abstracts

### Abstract 14

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**Abstract Title:** “Presenting symptoms and time to diagnosis for pediatric central nervous Tumors in Qatar: a report from pediatric neuro oncology service in Qatar.”

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# The Author(s) 2020

**Introduction:** There are no previous published reports on primary pediatric tumors of the central nervous system (CNS) in Qatar. We undertook this retrospective cohort study to review the diagnosis of CNS tumors in children in Qatar to analyse the presentation characteristics including symptoms, referral pathways, and time to diagnosis.

**Methods:** All children registered with Pediatric Neuro-Oncology service (PNOS) were included in the study. Data from the time of diagnosis (October 2007 to February 2020) were reviewed retrospectively. Presenting symptoms were recorded and pre-diagnosis symptom interval (PSI) was calculated from the onset of the first symptom to the date of diagnostic imaging.

**Results:** Of the 61 children registered with PNOS during the study period, 51 were included in the final analysis. Ten children were excluded because they were either diagnosed outside Qatar (n = 7) or were asymptomatic at the time of diagnosis (n = 3). The median age was 45 (range 1–171) months. Common tumor types included low-grade glioma (LGG) (47.1%) and medulloblastoma/primitive neuroectodermal tumors (PNET) (23.5%). Nine children had an underlying neurocutaneous syndrome. Thirty-eight patients (74.5%) had at least one previous contact with healthcare (HC) professional, but 27 (52%) were still diagnosed through the emergency department (ED).

Presenting symptoms included headache, vomiting (36.2%), oculo-visual symptoms (20.6%), motor weakness (18.9%), seizures, ataxia (17.2% each), irritability, cranial nerve palsies (12% each), and endocrine symptoms (10.3%). Median PSI was 28 days (range 1–845days) for all CNS tumors. Longest PSI was seen with germ cell tumors (median 146 days), supratentorial location (39 days), and age above 3 years (30 days). Tumor characteristics of biological behaviour (high-grade tumor) and location (infratentorial) were significantly associated with shorter PSI, as were presenting symptoms of ataxia, head tilt, and altered consciousness.

**Conclusions:** Although overall diagnostic times were acceptable, some tumor types were diagnosed after a significant delay. The awareness campaign, such as the “Head Smart” campaign in the United Kingdom (UK), can improve diagnostic times in Qatar.

Further research is required to better understand the reasons for the delay.

**Key words:** child. Brain. Delay in diagnosis.

## Abstract 15

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**Abstract Title:** "Imaging features surrogates for molecular subgroups of Medulloblastoma-An initial experience benefitting from twinning with Sick Kids, Toronto."

**Abstract Theme:** Pediatric Neuro Oncology

**Introduction:** Recent advances in genomics have led to the division of medulloblastomas into 4 groups which are wingless (WNT), sonic hedgehog (SHH), group 3, and group 4. This has shown potential for improved risk stratification as different subgroups show different clinical behaviors and may be subjected to subgroup-specific treatment as well. Recent literature has shown that different subgroups of medulloblastoma show different subgroup-specific imaging features.

**Methodology:** We retrospectively reviewed the imaging of all patients with biopsy proven.

medulloblastoma from the period of April 2015 till Dec 2019 which were sent for genomic testing to Toronto, Canada, in collaboration with Sick Kids Foundation. A total of 17 cases were included. However, cases with inconclusive genomic testing and unavailable pre surgical imaging were excluded. Cases were reviewed for important radiological features such as location of the lesion, contrast enhancement and margins of the lesion.

**Results:** In our initial review, we found 6 cases of group 4 tumors, 3 cases of SHH and 1 case of WNT. No case from group 3 was found in our review.

Group 4 tumors were mainly found in the midline, have well defined margins and show heterogenous post contrast enhancement with little to no peritumoral edema. The SHH subgroup of Medulloblastoma mainly lies lateral to the midline, in the cerebellar hemisphere. However, these tumors also show heterogenous post contrast enhancement, well defined margins and mild perilesional edema.

Only a single case of Wingless tumors was reviewed which was midline in location however shows homogenous post contrast enhancement.

**Conclusion:** In conclusion, Medulloblastoma depict characteristic imaging features according to their genomic subgroups. Location being the most distinctive feature differentiating SHH tumors from Group 4 and WNT sub-types. WNT and Group 4 subtypes, though midline tumors, can be differentiated based on their enhancement pattern.

**Key words:** Medulloblastoma, MRI, Genomic testing"

## **Abstract 16**

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**Abstract Title:** “Different morphological and histopathological features of pilocytic astrocytoma involving various CNS locations –Series of cases.”

**Abstract Theme:** Pediatric Neuro Oncology

### **Background:**

Pilocytic astrocytoma is the most common CNS tumor in children and adolescence and accounts for 15.5% of all cases. According to WHO classification, it is considered as low grade, WHO grade I CNS tumor. It can arise from anywhere though out central nervous, however it most commonly arises from cerebellum, with second most common location of optic chiasm. Other less common location includes brain stem, hypothalamus, cerebral hemispheres and spinal cord. Imaging features typically consist of well-defined usually cystic lesion with solid enhancing nodular component.

### **Discussion of Cases:**

In this study, we describe the five cases of histopathologically proven pilocytic astrocytoma of different regions of central nervous system. Three of these are seen in children below 16 years and two cases are of adult of age 22 and 23 respectively. Histopathology of three of these cases is also described. Locations of these fives lesions includes cerebellum, suprasellar region and optic chiasm, cerebral hemisphere, brain stem and fourth ventricle.

**Conclusion:** Pilocytic astrocytoma has a wide spectrum of neuroradiological presentations. Besides its classical appearance as low-grade glioma, a more atypical presentation makes the diagnosis challenging. The best method to achieve the pre-operative diagnosis is the combination of morphological and non-morphological MR features as well as site base approach of this tumor.

## Abstract 17

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**Abstract Title:** "Grading and immunohistochemistry of supratentorial ependymomas."

**Abstract Theme:** Pediatric Neuro Oncology

**Objective:** Supratentorial ependymoma is an uncommon tumor of the central nervous system that is challenging to classify. We described the histology and immunohistochemistry of supratentorial ependymomas in patients managed at the King Hussein Cancer Center.

**Methods:** We performed a retrospective chart review of cases between January 2003 and December 2013 and retrieved the corresponding tumor samples for histologic and immunohistochemical evaluation. We graded the tumors according to both the Marseille grading system and 2016 WHO classification and enumerated the presence or absence of three features of anaplasia, namely active mitosis, microvascular proliferation, and necrosis. We measured associations between tumor grade and the immunohistochemical markers using the N – 1 corrected Pearson's  $\chi^2$  test.

**Results:** We included 18 patients in the final analysis; 10 (55.6%) were children or adolescents and eight (44.4%) were adults. The results of Marseille and WHO grading were completely concordant. Nine cases (50.0%) were classic ependymoma (Marseille low-grade; WHO grade II) and nine (50.0%) were anaplastic ependymoma (Marseille high-grade; WHO grade III). None of the cases of classic ependymoma had more than one feature of anaplasia. All nine cases of anaplastic ependymoma had at least two features of anaplasia, and seven cases (77.8%) had all three features. The median Ki-67 index was 17.5% (range, 1.0–70.0%). Ki-67 expression was statistically significantly associated with tumor grade using the median as the cutoff value ( $p = .003$ ). None of the other immunohistochemical markers (GFAP, EMA, CD99, p53, cyclin D1, nestin, and H3K27me3) were associated with tumor grade.

**Conclusion:** Ki-67 expression discriminates between low- and high-grade supratentorial ependymal tumors. In addition, we provide evidence for the cross-reliability of the Marseille and WHO grading systems.

**Keywords:** Ependymoma; Immunohistochemistry; Supratentorial Neoplasms"

## Abstract 18

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**Abstract Title:** “Glioblastoma in adolescents and young adults: an age-based comparative study from Jordan over a 17-year period.”

**Abstract Theme:** Pediatric Neuro Oncology

**Objective:** Glioblastoma (GBM) is the most common primary brain tumor. Epidemiologic studies on GBM are rarely reported in the special age group of adolescents and young adults (AYA) compared with other age groups. We aim to present data on GBM in Jordan, with a focus on the AYA age, including the incidence, gender, location, and outcome, including long-term survival.

**Methods:** Data on GBM were requested from the Jordan Cancer Registry and statistical analysis was performed. All data were retrospective and anonymized.

**Results:** Eight hundred GBM cases were analyzed from 2000 to 2016, including 505 males (63.1%). Males outnumbered females across the studied years ( $p < 0.001$ ). There were 49 pediatric (0–14 years, 6.1%), 125 AYA (15–39 years, 15.6%), 358 adult (40–59 years, 44.8%) and 268 elderly (60+ years, 33.5%) cases. Supratentorial location predominated across all age groups ( $p < 0.001$ ). The preponderance of males and supratentorial tumors remained across the AYA age group in comparison to others. The median overall survival was 23.61 months. The AYA age group had a better outcome in comparison to the adults/elderly age group ( $p < 0.001$ ). Long-term survival was more common in the AYA age group ( $p = 0.021$ ).

**Conclusion:** This is the first comparative epidemiologic study of GBM in Jordan focusing on the AYA age group. The AYA age group appears to be associated with a better outcome compared with older age groups.

**Keywords:** Adolescent; Glioblastoma; Survival Analysis; Young Adult"

## Abstract 19

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**Abstract Title:** “Clinical profile of Ependymoma in children: single center study

**Abstract Theme:** Pediatric Neuro Oncology.”

**TITLE:** - “Clinical profile of Ependymoma in children: Single Center Study.”

Syed Habib Ahmed, Uzma Imam, Fahad Mumtaz, Marvi

**Introduction:** “Ependymoma most commonly appears in the fourth ventricle in children. It is the most common brain tumor in children and accounts for 5 to 10% of all pediatric brain tumors.”

**Objectives:** To evaluate the demographic, clinical, pathological features and treatment outcome of children with Ependymoma.

**Materials and Methods:** We reviewed the charts of all children with Ependymoma presented to oncology department, children cancer center (Child Aid Association) at National Institute of Child Health Karachi during October 2018 to September 2020. We collected data including age, gender, clinical features, type of histology and treatment outcome during this study period. Analysis was performed on SPSS version 20.

**Result:** Total 15 patients were included in our study. Age ranges between 2 to 15 years with mean age of 7 years. Male were n=13 (86.6%) and the female n= 2 (13.3%) with M:F ratio 6.5:1. Most of the patient came from interior Sindh which was n= 6 (40%). Headache is the major symptom n= 13(86.6% ) vomiting n=12 (80%), fever n= 4(26.6%), In majority of the patients Ependymoma located in posterior fossa n=12 (80%), most common histological type Anaplastic Ependymoma n=9 (60%). Two patient had metastatic disease 13.3%. Subtotal resection was performed in n= 8 (53.3% patients). 9 (60%) patients got conventional radiation with total radiation dose of 54.4 Gy and n=3 (20%) patients got chemotherapy before surgery. Three patients refused treatment. Among those 12 patients who received treatment 2 patient were put on palliative care, out of the 10 patients who received active treatment n=7 (70%) are alive with stable disease and n=3 (30%) died within 6 to 8 months after treatment.

**Conclusion:** In this study we found most of the patient had delayed presentation and came with advance stage disease. Mainly subtotal resection was done. Only with awareness in the community, health care professionals and multi-disciplinary team management, the survival outcome of these children will improve.

**Keywords:** Brain Tumor, Ependymoma, Children.

## Abstract 20

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**Abstract Title:** “Gray matter structural plasticity in patients with basal ganglia germ cell tumors: A voxel-based morphometry study.”

Basal ganglia; Germ cell tumors; Voxel-based morphometry; Magnetic resonance imaging

**Objective:** To assess structural brain plasticity in the presence of unilateral basal ganglia germ cell tumors (BGGCTs).

**Methods:** To detect changes in the gray matter volume of the whole brain, we applied voxel-based morphometry (VBM) to structural magnetic resonance imaging (MRI) scans. We compared a sample of 41 patients with BGGCTs in the left (BasalG\_L group;  $n = 22$ ) or right basal ganglia (BasalG\_R group;  $n = 19$ ) and a sample of 16 patients with germ cell tumors (GCTs) in pineal or suprasellar regions to 16 age-matched normal controls (NCs) using a two-sample t test, correcting all results for family-wise-errors. Results In patients with left BGGCTs, whole brain VBM analysis revealed a large cluster of voxels reflecting an increase in gray matter volume in the left para hippocampal region ( $k = 529$  voxels,  $T = 4.18$ ,  $p < 0.01$ ) and a decrease in volume in the left thalamus ( $k = 527$  voxels,  $T = -4.88$ ,  $p < 0.01$ ). At the same time, in patients with right BGGCTs, a cluster of voxels revealed an increase in gray matter volume in the right middle cingulate cortex (rMCC) ( $k = 172$  voxels,  $T = 3.96$ ,  $p < 0.01$ ), and a decrease in gray matter volume in the right inferior frontal gyrus (rIFG), pars opercular ( $k = 495$  voxels,  $T = -4.29$ ,  $p < 0.01$ ). Furthermore, there were no significant differences in the gray matter volume between groups of patients with GCTs in the pineal or suprasellar regions and NCs on a two-sample t test.

**Conclusions:** These results demonstrate that slow growing but destructive BGGCTs markedly and asymmetrically atrophy gray matter in specific brain regions and show compensatory plasticity in both cerebral hemispheres. Our findings show that whole cerebral adaptations in patients with BGGCTs may perhaps be the physiological basis for the high levels of functional compensation and partially explain the relationship between gray matter remodeling and their observed cognitive disturbances.

## Abstract 21

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**Abstract Title:** “Radiotherapy practice for Pediatric CNS tumors: Eexperience of tertiary care hospital.”

**Abstract Theme:** Pediatric Neuro Oncology

**Backgrounds and Aims:** The burden of pediatric CNS tumors requiring radiotherapy (RT) in developing countries is poorly understood and there is limited data available of radiotherapy experience for pediatric CNS tumors in our part of the world. This study aims to improve collaboration between radiation oncologist and other specialists by reviewing pediatric CNS radiotherapy practices at the Aga Khan University Hospital, Karachi through our experience.

**Method:** Hospital information management system (HIMS), cancer registry and radiation oncology record system were searched to identify children aged up to 18 years of age and received radiation therapy to CNS on the pediatric protocol. Data was collected for age, diagnosis, site, dose and other parameters related to radiation therapy.

**Results:** We identified 88 patients treated on pediatric protocol for primary CNS tumors from January 2009 till December 2019. Mean age was 11.3 years. There were 60 (68%) males and 28 (32%) female patients. All the patients were discussed in tumor board before being referred for radiation treatment. General anesthesia was used to treat 18 (20%) children. A total of 51 (58%) children received RT to the partial brain, 1 patient received whole-brain RT while 36(40%) children received CSI (craniospinal irradiation). Histologies associated with patients receiving CSI were medulloblastoma 29 (80%), ependymoma 3(10%), pineoblastoma 2(5%) and PNET 2 (5%). Partial brain RT where was delivered to Histologies including diffuse astrocytic and oligodendroglial glioma 41(72%), ependymoma 7(20%), other astrocytic tumors 2(5%), and germ cell tumors 1(3%). One patient with CNS lymphoma received whole-brain RT. Total RT dose was 59.4Gy with dose range of CSI in between 50.4-59.4 Gy. Most of the patients had their systemic treatment at our hospital or the other major pediatric oncology centre.

**Conclusion:** Radiation therapy has been and will continue to be a critical component of the multidisciplinary approach required in the care of children with CNS tumors. Further building upon this database will help find outcome related to disease control and late effects in relation to the extent of disease, dose of radiation therapy and treatment site.



## Abstract 22

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**Abstract Title:** “Pediatric Brainstem Glioma: A Tunisian single-center experience.”

**Abstract Theme:** Pediatric Neuro Oncology

**Introduction:** Brainstem gliomas are relatively rare tumors of the central nervous system. They have various presentations and clinical courses. The aim of this study was to analyze the clinical profile and therapeutic outcomes of these tumors.

**Key words:** brainstem glioma, radiotherapy, prognosis

**Methods:** A retrospective study of 8 patients diagnosed with brainstem glioma in the department of radiotherapy of Farhat Hached hospital between January 2005 and December 2018.

**Results:** The median age was 6 years (4-13 years). There were 4 girls and 4 boys. The diagnosis was made urgently in 3 patients, after 1 month in 4 patients and 2 months in one patient. The tumor was pontine for all the patients. The most common manifestations were ataxia, half body paralysis and headaches. Symptoms related to cranial nerve dysfunction were present in all patients. Diagnosis was confirmed by characteristic imaging on Magnetic Resonance Imaging. The average tumor size was 43 mm (6-55 mm). Contrast enhancement was commonly not prominent. The lesion appeared as hypo intense on T1 weighted images and hyper-intense on T2 weighted images. On spectroscopy, marked elevation of choline with reduced N-Acetylaspartic acid was observed. Hydrocephalus was present in 4 patients. Three of them had ventriculoperitoneal shunt placement. Only 5 patients were treated with exclusive conformal radiotherapy (RT) while the others died before treatment. The median RT dose was 54 Grays (Gy) (32,4-54 Gy). One patient died under treatment. A partial response was observed in three patients. There was one case of clinical and radiological progression treated by salvage radiotherapy at dose of 18 Gy. After a median follow up of 10 months (4-42 months), 7 patients died, and 1 patient was clinically and radiologically stable.

**Conclusion:** Brainstem tumors are aggressive with poor prognosis due to challenges in their management.

## Abstract 23

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**Abstract Title:** "Pediatric brain tumors: about 53 cases."

**Abstract Theme:** Pediatric Neuro Oncology

**"Introduction:** Brain tumors are the second most common cancer of children. Although children with brain tumors have a better prognosis than adults, treatment presents special problems, including possible damage to the developing brain. This study reviews epidemiological, clinical, therapeutic aspects of pediatric brain tumors.

**Patients and methods:** We reviewed 53 pediatric patients (less than 16 years old) with brain tumors, referred to our Department of radiation oncology, Sousse Farhat Hached Hospital-Tunisia, during the period from January 1995 to December 2018. Only tumors for which pathologic specimens were available were included.

**Results:** The mean age was 8 years with male prevalence (sex ratio =1,17).

Pathological examination showed that the most common tumors were glial tumors (60.3%), medulloblastomas (34%), craniopharyngiomas (3.8%) and germinal cell tumors (1.9%). While most tumors were more predominant in males, astrocytic tumors and craniopharyngiomas were more predominant in females. The most common symptoms were intracranial hypertension (43.30%) and cerebellar syndrome (30.18%). All our patients had brain imaging: 11.32% CT scan versus 88.68% MRI. The therapeutic component includes surgery, chemotherapy and radiotherapy: 75,5% had surgery ; 81,6% had radiotherapy and only 23% had chemotherapy. Radiation therapy doses varied from 20Gy to 60Gy The evolution was variable: 43.46% complete remission ,39.6% Dead ,9.4% relapse ,7.54% stability.

**Conclusions:** A small body of evidence has emerged, highlighting the marked heterogeneity of results, limiting our ability to draw solid conclusions. At this point, considerable progress has been made in the outcomes of certain tumors, prognosis in other childhood brain tumor types is poor.

**Key word:** Pediatric, Brain tumors, radiotherapy, Chemotherapy.

## Abstract 24

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**Abstract Title:** "Management of Pediatric Medulloblastoma in Sousse: about 26 cases."

**Abstract Theme:** Pediatric Neuro Oncology

**"Introduction:** Medulloblastoma (MD) is the most common malignant brain tumor in children, comprising 40% of all childhood posterior fossa (PF) tumors. It is potentially curable, and prognosis depends of the like hood of disseminated disease at the time of diagnosis.

We retrospectively reviewed 26 patients treated for MD in the radiation oncology department and the medical oncology department Farhat Hached Hospital, Sousse, Tunisia between 1996 and 2020.

**Results:** Median age at diagnosis was 7.8 years(1-15)with a sex ratio of 1. The main reason for consultation was intracranial hypertension syndrome in 13 patients with an average consultation time of 2months(1-5). Cerebral spinal fluid involvement was detected in 5 children. Overall, 25 patients had gross or near-total surgical resection. It was incomplete only in 5 cases. Histologically, MD subtypes were distributed as follows: 4 desmoplastic, 4 classic, 2 with extensive nodularity and 1 anaplastic. 9 infants were categorized as having high-risk disease. Radiotherapy(RT) was indicated to all patients but only delivered to 24 at a dose of 23.4 to 36Gy(1.8Gy/fraction) to the craniospinal axis with a complement in the PF at a dose of 54 to 56Gy. 21 patients received adjuvant Chemotherapy(CT) associating essentially VP16-Carboplatin-Vincristine-cyclophosphamide(50%) and 8 had concomitant RT-CT based on Vincristine. 4 patients kept neurological sequelae of which 2 had gait impairment, 1 neurocognitive disorders and 1 kept akinetic mutism.

Eleven relapses (42.3%) were observed after median time of 11 months (3-76) mostly localized in the PF (87.5%). Reirradiation was performed in one case at a dose of 15 Gy in the PF. After a mean follow-up of 39 months, 11 deaths were noted, 13 children were in full remission and 2 lost to follow up. Median overall survival was 22.5 months (2-248).

**Conclusions:** In accordance with the literature data, our study suggest a potential survival benefit from multimodal treatment in pediatric MD but recurrence remains high with a rate exceeding 30%. The challenge of MD treatment is how to achieve a maximum of benefits with minimum of toxicities. Such measures include a multidisciplinary approach.

**Key words:** Medulloblastoma/Radiotherapy/Chemotherapy.

## Abstract 25

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**Abstract Title:** "Pediatric intraspinal lesions: Radiological spectrum of ependymoma vs astrocytoma."

**Abstract Theme:** Pediatric Neuro Oncology

**"Introduction:** Intramedullary spinal lesions are a rare entity, accounting for less than 10 percent of all pediatric central nervous system tumors (1). Ependymomas and astrocytoma comprise almost 90 % of all intramedullary spinal lesions in children (2) They are slow growing tumors but their prognosis is mostly guarded (3). Previous studies have reported that pediatric intramedullary spinal lesions appear differently as compared to the adults (4). MRI is not only non-invasive but currently it is the modality of choice as it can provide excellent characterization and localization of spinal lesions. (5)

**Objective:** To determine the imaging features that differentiates between intramedullary ependymoma vs astrocytoma on MRI in pediatric patients.

**Material and Methods:** A retrospective analysis was performed of 22 biopsy proven cases of intramedullary ependymoma and astrocytoma at Aga khan Hospital from year 2008 to 2020 of pediatric patients (age < 18). Preoperative MRI images were reviewed, and data was collected from a radiology search engine and picture archiving and communication system (PACS) .2 case was excluded from the study due to incomplete data availability.

Data regarding the tumor location, size, margins, enhancement, signal intensity, mass effect, syringohydromyelia, cystic component, hemosiderin cap sign and hemorrhage was collected.

**Results:** Out of 22 cases, 13 cases were of ependymoma and 7 cases were of astrocytoma.

Most of the lesions were found to involve cervical spine (75%), followed by dorsal spine (25%). Older age, solid component and intense enhancement is seen more frequently in ependymoma. Hemorrhage and hemosiderin cap sign were found in none of the case. Syringohydromyelia was found only in ependymoma in the collected sample.

**Conclusion:** Out of multiple characteristic features, syringohydromyelia is found to be the distinguishing radiological feature on MRI in between ependymoma vs astrocytoma.

## Abstract 26

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**Title:** "Embryonal Tumor with Multilayered Rosettes (ETMR)."

**Theme:** Pediatric Neuro Oncology

**Background & Objectives:** The 2016 update of the WHO Classification of Tumors of the CNS has redefined a number of tumors. Embryonal Tumor with Multilayered Rosettes is a recently described pathological entity. They are rare tumor affecting young children. Prognosis is extremely poor.

**Methods:** We retrieved and reviewed H&E stained microscopy glass slides of 13 ETMR cases with history of headache, vomiting and pain. These cases were treated at different institutes and diagnosed at our institute between March 2008 and July 2019. Morphologic features such as presence of papillary/tubular-trabecular architecture, surface blebs, external blebs, primitive cell component, neuronal, glial and mesenchymal differentiation were assessed.

**Results:** Patients' age ranged from 8 months to 10 years with median of 3.63 years. Female to Male ratio was 3:2. Brain was the most commonly involved site.

Most of the tumors were received in multiple pieces. All the cases showed features of malignancy. Follow-up of 6 patients could be attained and 3 were found to have expired due to some undiagnosed disease rest were all healthy with 2 weeks follow up history. Treatment of all patients was excision alone with few patients who received radiotherapy.

Microscopically, as presence of papillary/tubular-trabecular architecture, surface blebs, external blebs, primitive cell component, neuronal, glial and mesenchymal differentiation was observed in all cases.

**Conclusion:** ETMR are a distinct entity with majority of them behaving aggressively even after being treated by local excision. Although the WHO currently recognizes 3 distinct histopathological entities-embryonal tumor with abundant neuropil and true rosettes, ependymoblastoma, & medulloepithelioma. Recent studies indicate that these tumors have a common molecular profile and clinical course and that they are now classified as a single entity."

## **Abstract 27**

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**Abstract Title:** "Constitutional mismatch repair Defect; A case series."

**Theme:** Pediatric Neuro Oncology

**Other Theme:** Inherited Cancer Predisposition Syndrome

**"Background:** Constitutional mismatch repair deficiency is a distinct childhood cancer predisposition syndrome showing autosomal recessive pattern of inheritance. This rarely seen disorder has been reported to manifest in infants or young adulthood with an incidence of one per million patients.

DNA Mismatch Repair is a system for detection and subsequent repair of errors in newly synthesized DNA during DNA replication. These errors may result from erroneous deletions or faulty incorporation of bases. MMR system comprises of four genes named as MLH1, MSH2, MSH6 and PMS2.6. Heterozygous or monoallelic germline mutations of these genes leads to autosomal dominant syndrome Lynch Syndrome while homozygous or biallelic mutations of any of these aforementioned genes may result in a cancer predisposition syndrome called CMMRD.

### **Case series of CMMRD.**

**Case no.1:** 11years old male was diagnosed as Glioblastoma Multiforme (GBM), IDH- wild type WHO grade IV. Histopathology revealed immune staining retained for MLH1 and PMS2. He underwent gross total resection followed by radiotherapy. Meanwhile he developed shortness of breath and further work up revealed T cell Leukemia. His elder sibling was diagnosed as T cell lymphoma two years back but succumbed to death before receiving any treatment. The paternal uncle of these patients lost 5 children owing to malignancies.

**Case no.2:** A 6 years old female child was diagnosed as T cell lymphoblastic lymphoma. Her brother died of GBM 2years back. On further testing, histopathology of mediastinal mass revealed retained immune staining for MLH1, MSH2 and MSH6.

**Case no.3:** An eighteen years old female diagnosed with Glioblastoma Multiforme (GBM), no significant family history, no café au lait spots. On further testing histopathology of tumor revealed retained immune staining for MLH1, PMS2, MSH2 and MSH6.

**Conclusion:** CMMRD is a highly malignant syndrome with poor prognosis in our setting. Early diagnosis for abnormal expression of genes can guide to start early surveillance in these children."

## Picture Poster Abstracts

### Abstract 28

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**Abstract Title:** “Acute morbidity and mortality analysis of COVID-19 positive children on cancer treatment: Experience from a lower middle-income country”

**Abstract Theme:** Pediatric Neuro Oncology

**Other Theme:** Covid-19 in children with cancer

**Purpose:** 1. To determine overall morbidity and mortality of COVID-19 infection in children with cancer treatment

2. To describe clinical characteristics of COVID -19 infection in children on active treatment of cancer.

**Materials and Methods:** Retrospective analysis of impact of COVID-19 infection on children who were on active cancer treatment from 1/04/2020 to 31/07/20. Data fields included demographic variables, diagnosis, treatment category, clinical feature with COVID-19 infection, duration of hospital stay, ICU admission, impact on treatment and outcome were noted.

**Results:** During the study period of 4 months (April to July 2020) a total of 19 pediatric oncology patients tested positive.

In terms of presenting symptoms 5 patients were asymptomatic and were diagnosed as part of pre-surgery or pre-chemotherapy testing, 9 patients had fever in combination with cough, sore throat, body aches or diarrhea and two patients presented with sore throat, rash, cough and body aches without a fever. Two of the patients had a concurrent infection. In terms of radiologic finding, CXR was not done in 8 patients and 3 showed a variety of findings including diffuse infiltrates and patchy air space disease. Ten patients required hospitalization. Only 4 of 19 patients required oxygen and there were two intensive care admissions. The median duration of hospitalization was 5 days with a range from 1 to 30 days. There was one mortality due to COVID 19 related pneumonia.

There were 4 patients in which there was not impact on cancer therapy for the rest there was an average delay in chemotherapy of 8 days (1-14 days) and surgery of 15 days (12 –20 days). Radiation was delayed for one patient.

**Conclusion:** The limited but accumulating evidence suggests that patients with cancer are at risk of COVID-19 infection, causing interruption in cancer treatment which can cause poor disease control.

## Abstract 29

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**Abstract Title:** “Primary central nervous system sarcoma in childhood: A case report of DICER1 variant CNS sarcoma diagnosis and treatment.”

**Abstract Theme:** Pediatric Neuro Oncology

Primary central nervous system sarcoma in childhood: A case report of DICER1 variant CNS sarcoma diagnosis and treatment.

**Introduction:** Central nervous system (CNS) sarcomas are exceptionally rare in pediatric patients, historically associated with an exceptionally poor prognosis. Here, the authors present a novel case of DICER 1 associated CNS sarcoma. Its clinical presentation, diagnosis and treatment aspects.

**Case Report:** A 16 years old girl presented in emergency department with history of headaches and vomiting for 10 days, diplopia for 4 days, altered level of consciousness for 12 hours. Patient had a single episode of generalized tonic-clonic seizures. She was drowsy with no eye opening or verbal response, no motor movement (on deep pain). Pupils were fixed. Cough, gag and corneal reflexes were weak present. Plantars were mute. Other systemic examination was unremarkable. MRI brain done immediately showed a large space occupying lesion in brain occupying frontal, parietal and temporal regions along with hemorrhage. Neuro navigation guided craniotomy and excision of lesion done. Immunohistochemistry and molecular analysis showed it to be DICER 1 associated CNS Sarcoma.

Oncogenic TP53 P.R158 H, DICER1 P.E 707 and DICER 1 p.E1813A mutations.

No MYCN amplification. KRAS amplification within a 5 Mb region at 12p12.1.DD1T3 and CDK4 amplification within a 4Mb region at 12q13.3 -q 14.1.MDM 2 amplification within a 1.5 Mb region at 12q15.Chemotherapy ICE protocol is started and well tolerated. Subsequent surveillance, a second look surgery and focal radiation in plan.

**Conclusion:** CNS sarcomas in pediatric age group are rare but behave as high-grade CNS tumors and may be associated with other benign or malignant conditions of body. We suggest early diagnosis prior to dissemination, complete surgical resection, systemic chemotherapy and focal radiation as an essential treatment goal in this rare disease.”



## Abstract 30

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**Abstract Title:** “Erdheim-Chester disease (ECD) a rare case of isolated CNS presentation.”

**Abstract Theme:** Pediatric Neuro Oncology.”

**Introduction:** Erdheim-Chester disease (ECD) is a rare non-Langerhans cell form of histiocytosis that can affect the central nervous system. ECD predominantly affects adults, and only a few pediatric cases have been reported till date. ECD symptoms depend on which organ(s) is involved, which varies with each patient. In this article, the authors describe the intricate diagnosis and treatment processes in this patient.

**Case Report:** A 14 years old boy presented with history of headache on and off for 2 years, fever on and off 1 year. He has been treated for migraine initially for around 2 years then developed 2 episodes of amnesia and was diagnosed and treated for epilepsy for a period of another year. When symptoms did not improved brain imaging was done that showed a heterogeneous enhancing left parietal extra-axial lesion. He underwent neuro navigation guided left posterior parietal craniotomy. Histopathology, at our institute favored it as Histiocytic Sarcoma. As part of our twinning with The Hospital of Sick Children, Toronto, Canada, we have sent the sample for second opinion and it was reported as Erdheim Chester Disease/ Extracutaneous Juvenile Xanthogranuloma with atypical features - Positive for IRF2BP2-NTRK1 fusion transcript. The presence of an NTRK fusion allowed us the use of targeted therapeutics. Post operatively remained well and discharged. He later on presented with sudden loss of vision after 6 weeks of surgical resection. A repeated imaging showed progression of the disease with multiple large intracranial lesions. CSF detailed report and cytology also showed CNS involvement. He was started on Chemotherapy Clidarabine therapy with four doses of Intrathecal (Methotrexate/Hydrocortisone). His condition improved significantly with significant improvement in vision and regression of disease on MRI Brain.

**Conclusion:** Erdheim-Chester disease (ECD) being rare in pediatric age group is difficult to diagnose yet increasingly molecular testing of biopsy is playing a part in an ECD diagnosis, as about 85% or more patients are seen to have a genetic mutation. A correct diagnosis relies upon findings of a biopsy of the affected tissue along with imaging studies. Early diagnosis and prompt treatment may change the outcome.”

## Abstract 31

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**Abstract Title:** “Field in field Technique with intrafractionally modulated junctions shift for craniospinal irradiation (CSI) planning with 3D-CRT at Ziauddin Hospital.”

**Abstract Theme:** Pediatric Neuro Oncology

**Keywords:** Craniospinal Irradiation, Field-in-field technique, Medulloblastoma

**Purpose:** To plan craniospinal irradiation with “field-in-field” (FIF) homogenization technique in combination with daily, intra-fractional modulation of the field junctions, to minimize the possibility of spinal cord overdose. Photon-based techniques for craniospinal irradiation (CSI) may result in dose inhomogeneity within the treatment volume and usually require a weekly manual shift of the field junctions to minimize the possibility of spinal cord overdose. Nowadays field-in-field technique is used to feather out the dose inhomogeneity caused by multiple fields. We have started using this technique after acquiring advanced technology machines in recent years.

**Methods and Materials:** 16 patients (2 adults, 14 children) treated with 3D-CRT for craniospinal irradiation were retrospectively chosen for this analysis. Most of the cases were of Medulloblastoma. These patients were planned and treated during 2016-2017. Contouring of Brain and Spine Cord and organ at risk were already done and planning done on Eclipse TM Treatment Planning System (Varian). All of these patients were planned Lateral cranio-cervical fields and posterior spinal fields were planned to use a forward-planned, FIF technique. Field junctions were automatically modulated and custom-weighted for maximal homogeneity within each treatment fraction. Dose volume histogram (DVH) was used for analysis of results. A corresponding plan without FIF technique was planned and maximum dose at the junction was noted for each patient with both plans and the readings were evaluated.

**Results:** Plan inhomogeneity improved with FIF technique. Planning with daily-modulated junction shifts provided consistent dose delivery during each fraction of treatment across the junctions. The maximum doses calculated at the junction were higher in the CSI plans without FIF compared to those with FIF technique.

**Conclusion:** This paper hence proves that FIF technique is better in planning craniospinal irradiation.”

## Abstract 32

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**Abstract Title:** “Pakistani Parents Experiences of Hope having Children with Cancer. A Qualitative Approach to Explore Parental Hope.”

**Abstract Theme:** Pediatric Neuro Oncology

**Other Theme:** Pediatric Oncology Nursing

**Background:** Different cultures and countries study the impact of family hope having children with cancer. Pakistani families have also associated many hopes regarding the care and treatment of their children having cancer.

**Objective:** This current study focuses on the impact and importance of hopefulness for parents of children with cancer, how hope relates to parents’ experience with the diagnosis, and the influence nurses and other health care professionals have on parents’ hope.

**Methodology:** Following an interview base format, 30 parents of children diagnosed with cancer were given the opportunity to express about hope through interview base questionnaire and asked 5 open-ended questions about hope. Answers were analyzed using content analysis. Parents’ adaptation to their child’s diagnosis was compared with answers to the hope questions.

**Results:** Parents defined hope as a knowing, belief, or wish regarding their child’s health. They emphasized the importance of hope over the course of their child’s treatment. Nurses increased parents’ hope by providing care to children and families, educating parents, and by connecting with and providing a positive viewpoint for families. Most parents felt there was nothing staff and nurses did to decrease their hope.

**Conclusion:** Understanding parents’ experiences validates the quality care and connections we make with children and families and encourages us to consider the effects of our interactions. This underscores the importance of education and support as a means of instilling hope in parents, who are valued, critical members of their child’s health care team.

**Keywords:** parents, children, cancer, hope"

## Abstract 33

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**Abstract Title:** “Neurofibromatosis type 1 and brain tumors.”

**Abstract Theme:** Pediatric Neuro Oncology

**Introduction:** Neurofibromatosis type 1 (NF1) is an autosomal dominant inherited disorder characterized by multisystemic symptoms including a predisposition to brain tumors particularly optic nerve gliomas.

**Methods:** A retrospective study was conducted at the Pediatric Oncology Unit in Tunis Children's Hospital from 2016 to 2020. It includes children diagnosed with a brain tumor associated with NF1 according to the National Institutes of Health.

**Results:** 5 cases were collected. The sex ratio was 0.4. No family history of NF1 was noted. In three patients, the diagnosis of NF1 was made at the same time as the brain tumor while in 2 patients, the tumor appears few years after the diagnosis. The diagnosis of NF1 was based on clinical findings: café au lait spots, axillary and inguinal freckling. Molecular genetic testing for analysis of NF1 locus was performed in only one patient. The mean age at diagnosis of the brain tumor was 9.8 years (4-14 years). The circumstances of brain tumor discovery were hemiparesis (2), loss of visual acuity (1), nystagmus (1), strabismus (1) and exophthalmia (1). 2 patients had glioblastoma treated by surgical resection followed by chemotherapy concomitant to radiotherapy. Three patients having optic nerve low grade gliomas were treated by chemotherapy according to the SIOP-LGG 2004 protocol. The sequelae after the end of chemotherapy were blindness (1), visual impairment (1) and ataxia (1). One patient having glioblastoma died by progression of the disease while the 4 other patients are alive with a stable residual tumor.

**Conclusion:** NF1 is one of the most frequent genetic diseases. Annual physical examination with ophthalmologic examination and cerebral MRI is imperative to detect early brain tumors (generally low-grade gliomas).

## **Abstract 34**

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**Abstract Title:** "Case Report 1.docx 1 / 6 Non-Hodgkin Lymphoma and Colorectal carcinoma: Metachronous occurrence in a patient with underlying DNA mismatch repair syndrome."

**Abstract Theme:** Pediatric Neuro Oncology

**Objective:** Constitutional mismatch repair deficiency (CMMRD) is an autosomal recessive disorder caused by biallelic mutations in DNA mismatch repair genes. These patients have clinical stigmata of neurofibromatosis 1(NF-1) with childhood onset of hematological malignancies, high grade gliomas and colorectal-cancers.

**Methods:** We present a case of non-Hodgkin's lymphoma (NHL) who later on developed adenocarcinoma colon at age of 11 years with significant family history of glioblastoma in elder brother and colonic cancer in mother. Results: This is first case of CMMRD in Pakistan who developed colonic neoplasm at age of 11 years. Nearly 150 patients of CMMRD have been reported worldwide.

**Conclusion:** Our case adds to the literature that colorectal cancer can occur in these patients during early adolescent age. Sibling screening and subsequent genetic counselling is important in countries like us where consanguinity is much prevalent.

**Keywords:** Colorectal Neoplasms, Constitutional mismatch repair deficiency, DNA Mismatch Repair, Lymphoma, non-Hodgkin's, Lynch syndrome"

## Abstract 35

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**Abstract Title:** "Low-grade gliomas in children: A single Tunisian experience."

**Abstract Theme:** Pediatric Neuro Oncology

**Introduction:** Central nervous system (CNS) tumors are the most prevalent malignancies in children. Gliomas account for 50% of all CNS malignancies. In low-grade gliomas (LGG), chemotherapy is the mainstream of treatment.

**Patients and Methods:** This study is a retrospective analysis of children with LGG treated in the Children's Hospital of Tunis from 2008 to 2020. The aim is to analyze the objective response to chemotherapy. Patients without histological diagnosis were included when the clinical and radiological features were consistent with LGG.

**Results:** 15 cases were collected. The sex ratio was 0.46. The mean age at diagnosis was 5.7 years. Circumstances of discovery were exophthalmia (4), visual impairment (4), nystagmus (4), strabismus (2), gait disorders (2) and back pain (1). Neurofibromatosis type 1 was associated in 3 cases. The gliomas were optic in 12 cases and intramedullary in 3 cases. Biopsy done in 6 patients was complicated by blindness in one case. Surgery performed in 5 patients was complicated by blindness and visual field amputation in 4 cases. 9 patients were treated by chemotherapy according to the SIOP-LGG 2004 protocol. Abstention was indicated in 2 patients. Sequelae were reported in 11 cases: blindness (3), static disorders (3), visual field amputation (2), sphincter disorders (2) and early puberty (1). All patients are alive with a regression of more than 30% of tumor in 8 patients; 2 patients relapsed. One patient is still under observation.

**Conclusion:** The diagnosis of LGG can be based on clinical and radiological data. Chemotherapy is preferred to radiotherapy because of the vascular, endocrine and neurocognitive risks. It has shown its effectiveness in stabilizing the disease."

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**Abstract Title:** "The Esthesioneuroblastoma: about two pediatric observations."

**Abstract Theme:** Pediatric Neuro Oncology

**Introduction:** Olfactory neuroblastoma ou esthesioneuroblastoma is a rare malignant neoplasm of the upper nasal tract arising from sensory neuroepithelial cells with neuroblastic differentiation. Fewer than 1000 cases worldwide have been published in the last 20 years. The treatment is based on surgery, chemotherapy and radiotherapy.

**Methods:** A retrospective study was conducted at the Pediatric Oncology Unit in Tunis Children's Hospital from 2016 to 2020 including children diagnosed with esthesioneuroblastoma.

**Results:** We reported the observations of two boys aged two and five years who were admitted for convergent strabismus complicated by rapid onset of bilateral blindness in one case and isolated voluminous cervical lymph node enlargement in the other case. The cerebral and facial MRI showed a voluminous central lesion of the sphenoid, compressing the optic chiasma and the optic nerves with cerebral extension in the first case and a right nasal ethmoidal process with endo cranial extension in the second case. The anatomopathologic exam of the biopsy showed a small blue cell proliferation expressing synaptophysin Chromogranin and PS100. The biological exam reported in one case hyponatremia which is believed to be secondary to inappropriate vasopressin secretion. The treatment consists of chemotherapy in the two cases. In the first case, the evolution was initially favorable after chemotherapy, but the patient died par tumoral progression a year after diagnosis. The second patient is under treatment.

**Conclusion:** Esthesioneuroblastoma is a rare malignancy in the pediatric population. Its development in confined structures explains its late diagnosis and frequent extension to neighboring organs. Actually, the development of new radiotherapy techniques and new anti-cancer drugs should improve the prognosis of this tumor and quality of life."

## Abstract 37

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**Abstract Title:** Re-irradiation of midline gliomas: about one case

**Abstract Theme:** Pediatric Neuro Oncology

**"Introduction:** Brain stem gliomas (BSG) account approximately for 10–20% of all childhood central nervous system tumors. They are classified into two groups: low grade BSGs and diffuse intrinsic pontine gliomas (DIPG). This second entity is newly termed diffuse midline glioma (DMG). Recent genomic outcomes have uncovered distinct mutation, the H3K27M, found in the majority of DMG. The prognosis remains poor with a 2 years survival of less than 10%.

**Methods:** we present a detailed description of DMG case in a 4 years old child treated in the radiation oncology department Farhat Hached Hospital, Sousse, Tunisia in 2019.

**Results:** A 4-year-old child complained of a 1-month history of progressive speech disorders, postural instability and gait impairment. Clinically, there were a cerebellar syndrome and bilateral nystagmus. Brain Magnetic resonance imaging (MRI) demonstrated infiltrating pontine lesion with bulbar and mesencephalic extension, it was 55\*50\*50mm sized process whose characteristics were in favor of pontine glioma. He had 3D conformal Radiotherapy (RT) at the dose of 54Gy in 30 fractions at the rate of 1.8Gy/session for the tumor with regression of neurological disturbances. The control MRI 3 months later revealed a tumor regression. After 6 months of follow-up, the MRI showed the pontine lesion which increased in size. The child was reradiated at the dose of 18Gy in 10 fractions. An improvement of neurological status was marked.

**Conclusion:** DMG remains an incurable disease. Only four in ten young people will be alive at one year after diagnosis. It is difficult to establish a standardized therapeutic strategy: owing to their poorly demarcated extension, DMG are generally inoperable. Irradiation is the only effective treatment delays transiently tumor progression and prolongs survival by a mean of 3–6 months. Hypofractionated RT and re-irradiation (rRT) at first progression have improved neurological symptoms with acceptable quality of life. Future clinical studies of rRT should further assess the benefit of this approach. A global coordination between pediatricians, neuro-oncologists and radiotherapist oncologists have to be placed meeting this challenge.

**Key words:** Diffuse Midline Glioma/Re-irradiation/Prognosis



## Abstract 38

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**Title:** Fetal Craniopharyngioma, A case report

**Theme:** Pediatric Neuro Oncology

**Other Theme:** Fetal/Neonatal Brain Tumor

**"Background:** Craniopharyngioma is a rare tumor of central nervous system representing <1% of all primary CNS tumors, but it constitutes the most common intracranial non-glial tumor in children. It usually arises from Rathke's pouch remnants, near the pituitary gland in suprasellar area and can extend to hypothalamus, optic chiasm, third ventricle and even to cranial nerves. It has bimodal age distribution, with first peak between 5-10 years and second between 55-60 years.

Craniopharyngioma rarely presents during fetal life and only few cases of antenatal diagnosis of craniopharyngioma have been reported. We present the case of a patient who got diagnosed with supra-sellar tumor on prenatal ultrasound and had surgical resection after birth.

**Case Report:** Our Patient was diagnosed on routine prenatal ultrasound at 26 weeks of gestation with 1.6 X 1.4 cm echogenic mass in inter-peduncular cistern raising suspicion of space occupying lesion in brain. The baby was delivered at 40 weeks by planned C-section, feeding well with unremarkable neurological examination. Between 6 to 9 months of age, parents noticed unusual increase in head circumference and inability to stand with support. He was brought to local hospital and initial MRI showed 5.2X 6.2 X 6.7 cm supra-sellar mass likely craniopharyngioma. Parents were advised to get neurosurgical consultation. Surgery was planned but due to some unavoidable circumstances got delayed for one more year. Patient had resection of the tumor in September 2020 at 2 years of age. Histopathology revealed nodules of plump anucleate squamous, ghost cells, wet keratin along with calcification and teeth like structure. Morphological features were suggestive of craniopharyngioma. Patient was stable after surgery and follow up MRI is planned.

**Conclusion:** Our patient is case of a rare presentation of an antenatal diagnosis of craniopharyngioma who had successful surgical resection. Surgical outcomes are poor due to large tumor size and are associated with high morbidity and mortality in the literature.

## Abstract 39

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**Abstract Title:** "Painful ophthalmoplegia revealing Burkitt's leukemia."

**Abstract Theme:** Pediatric Neuro Oncology

**"Introduction:** Burkitt's leukemia is a fast-growing malignancy of B cell typically associated with a rapid onset and progression of symptoms. The clinical presentation varies according to the initial site and the presence or no of metastases. We report a case of Burkitt lymphoma revealed by ophthalmoplegia and we propose to review literature.

**Observation:** We report a case of central nervous system Burkitt's lymphoma in a 11-year old boy presented with unilateral ophthalmoplegia. H developed weakness of the eye muscles of the right eye causing ptosis of the right lid. The third, fourth and sixth cranial nerves on this side were paralyzed on the right. The rest of the examination was without abnormalities. A cerebral MRI was performed in emergency showing a mass of the pituitary gland invading the right cavernous sinus. He developed few days later diffuse bone pain resistant to analgesic treatments and bicytopenia in the blood count numeration. A bone scan was performed and showed hyperfixation of the 2 tibias. The bone marrow aspiration confirmed the diagnosis of Burkitt's Leukemia. The patient responds well to chemotherapy according to LMB2001 in the high-risk group (stage IV). After 2 courses of chemotherapy, there was improvement of the right ptosis. At the end of treatment, the eye motility had returned to normal and cerebral MRI demonstrated absence of the previously cerebral tumour. The bone Marrow aspiration confirmed also the complete remission. After 18 months of follow-up, the child has progressed well, but neurological bladder persisted.

**Conclusion:** Ophthalmoplegia is a frequent reason for consultation in ophthalmology and pediatrics. However, when confronted with this problem, clinical examination should be very careful followed by investigations in order to distinguish infectious from neoplastic causes."

## Abstract 40

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**Abstract Title:** A Case of Infant MBEN indicated by Imaging and Literature Review

**Objective:** To summarize a case of infant medulloblastoma with extensive nodularity (MBEN) indicated by imaging, to help raise awareness of this disease. **Key Words:** infant; Medulloblastoma; medulloblastoma with extensive nodularity, MBEN; MDT; prognosis.

**Method:** The familial history, clinical manifestations, images and management were analyzed, and the literatures were reviewed. **Result:** This was a five-month old in-vitro fertilization baby boy with a history of asphyxia at birth and had neonatal jaundice. He had cancer family history. On routine head MRI scan after 3-month-old showed lesion in the cerebelli with obstructive hydrocephalus which indicated medulloblastoma. No management except mannitol to decrease the intracranial pressure until consultation to our hospital two months later. At that time, the mass had increased significantly. The parents give up the treatment in terms of the high risk of severe sequelae of blindness. On literature review, the imaging of this infant patient was mostly related to MBEN, for infant MBEN, the long-term outcomes reported by HIT2000 protocol showed 5-year non-progressing survival rate was up to 93% with 5-year overall survival rate 100%, while the long-term follow-up by COG ACNS1221 protocol indicated 2-year non-progressing survival rate was 92%, recurrence rate was 0.

**Conclusion:** MB is the most common embryonic malignant tumor of pediatric central nervous system brain tumor. The prognosis of children MB is unfavorable, and the survivals may have cognitive defect. While the prognosis of MBEN is favorable, with longer survival period and lower recurrence rate; it is the hopefully curable MB type. Therefore, diagnosis, treatment and the study of prognosis knowledge for infant medulloblastoma should be strengthened, so as to raise clinical doctors' cognition and improve the management for this disease. At the same time, multidisciplinary (MDT) collaboration should be stressed.

## **Abstract 41**

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**Abstract Title:** An adolescent case of intracranial germinoma with depression as the first manifestation

**Objective:** To Summarize a case of adolescent intracranial germinoma with the first clinical manifestation of depression and anorexia and to help clinicians aware the rare clinical manifestation for early diagnosis. **Methods** The medical history, imaging and treatment of the boy were summarized. **Results** This was a 14-year-old boy who was well until he visited the doctor in May 2020 for black mood, polydipsia and polyuria, headache and diplopia. He had been diagnosed depression since then. He did not improve after treatment with oral antidepressant (Sertraline, Venlafaxine). His symptom deteriorated and had suicidal tendency. The head MRI showed suprasellar lesion and had received surgery in Hunan Province. The pathology indicated germinoma and transferred to our hospital after one month delay due the shortage of bed in Hunan hospital. He received chemotherapy on COG ACNS 1123 protocol and got good response of the treatment not only for the imaging but also for the symptom of depression. It is the first report of adolescent with intracranial germinoma suffered from depression before diagnosis. **Conclusion** Psychiatric symptoms occur in about 20%-40% of intracranial tumors. The most common symptoms include polydipsia and polyuria, headache, impaired vision/visual field, and so on. No intracranial germinoma with the first manifestation of depression has been reported. Our case report can raise the awareness of intracranial germinoma in young children. Germinoma is sensitive to chemotherapy and radiotherapy. After treatment, the symptom will improve soon.

**Key words:** adolescent; intracranial germinoma; depression.

## Abstract 42

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**Title:** Choroid plexus carcinoma: A case report

**Theme:** Pediatric Neuro Oncology

**"Introduction:** Choroid plexus tumors are intraventricular tumors derived from choroid plexus epithelium. It's represents less than 1% of all brain tumors. These tumors affect children more often than adults. Choroid plexus carcinoma is the most serious form of these tumors according to the 2016 OMS classification. The objective is to discuss the epidemiological characteristics, diagnosis, treatment, and prognosis of this rare childhood tumor in the center of Tunisia.

**Observation:** We report the case of a 5-year-old child, without notable pathological history, admitted to the neurosurgery department for a symptomatology of rapidly progressive installation over 2 weeks of violent headache and vomiting. At the time of admission, his Glasgow Coma Scale (GCS) was 12/15. Papilledema was present bilaterally. Computed Tomography (CT) scan of brain revealed an expansive process in the occipital horn of the left lateral ventricle measuring 2 \* 1cm. The patient was operated on September 2019. Intraoperatively, the tumor was fragile and highly vascular. The boy underwent subtotal excision of the mass. Histopathology examination of the excised tumor was reported a choroid plexus carcinoma. Whole spinal MRI and CSF cytology were performed and both tests were negative for leptomeningeal seeding. Patient was planned for craniospinal irradiation to the primary lesion (50 Gy/30 fractions in 6 weeks). The child received 3 courses of adjuvant chemotherapy including Vincristine Etoposide, cyclophosphamide and carboplatin with severe side effects. The patient was followed-up with a magnetic resonance imaging (MRI) of the brain 3 months after chemotherapy which revealed a residual of tumor. So, we indicate a palliative chemotherapy including Temozolomide. Currently the child is doing well, but he still has asthenia. The radiological evaluation will be done after the 3ird chemotherapy cure.

**Conclusion:** Choroid plexus carcinoma is an uncommon tumor that usually occurs in the lateral and fourth ventricles. It is associated with a poor prognosis. The current accepted treatment for CPC is surgical excision with controversial use of adjuvant chemotherapy.

## **Abstract 43**

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**Title:** Pediatric Glioblastomas in the center of Tunisia

**Theme:** Pediatric Neuro Oncology

**"Introduction:** Glioblastoma in children accounts for no more than 3-5% of primary central nervous tumor in children ,in the absence of concrete evidence for adjuvant chemotherapy ,maximal surgical resection followed by adjuvant RT(in children >3years of age ) remains the current best treatment strategy for these tumors .

**Materials and Methods:** Between 1995 and 2018 we identified six pediatric glioblastomas confirmed by pathological examination in radiation oncology department, Farhat Hached Hospital Sousse Tunisia.

**Results:** The median age was 10 years (6-16 years) with ratio 2. The main reason for consultation was often intra-cranial high pressure. Right temporal locations were the most frequent. We describe 04 cases of glioblastoma; 01 glioblastoma with oligodendroglial component and 01 giantocellular glioblastoma.

All patients underwent surgical intervention. Two patients had gross total resection (GTR) and four patients had subtotal resection. After surgery, all patients received radiation therapy associated to adjuvant chemotherapy in three cases.

After a median follow-up time of 25 months, (5 days–120 months), 2 patients were in complete remission, 3 patients died, and 1 patient was lost of follow up.

The median overall survival was 14 months.

**Conclusion:** Pediatric glioblastoma is rare but distinctly different biological disease compared with adults ;prognosis in the majority of children is better than the adults which in turn may be explained by a different biological make up of these tumors ,the place of the genetic and molecular analysis is more than crucial to better understand this affection."

## Abstract 44

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**Title:** “Primary Central Nervous tumors in Pediatric Population: A retrospective study about 23 cases.”

**Theme:** Pediatric Neuro Oncology

**Introduction:** Primary Central nervous system (CNS) are the most common solid tumors in children and the second leading cause of cancer death.

They comprise a variety of tumor types, of which nearly 50% arise below the tentorium.

**Materials Method:** We retrospectively reviewed 23 patients diagnosed with primary CNS and treated in the medical oncology department Farhat Hached Hospital,Sousse,Tunisia.

**Results:** Median age at diagnosis was 6 years (0-14). A slight female predominance was noted (52.2%). the average time to consultation was 2 months (1-6) and the main reason for consultation was intracranial hypertension syndrome in 15 patients with or without motor dysfunction.

The tumor was infra-tentorial in 78.3% of cases and were dominated by tumors of the fourth ventricle and vermis (66.6%).

Six histological subtypes were found in our series: medulloblastoma(n=18), anaplastic astrocytoma(n=1), PNET(n=1), ependymoma (n=1), pineal blastoma (n=1), AT/RT (n=1).

Four patients have leptomeningeal spread disease at diagnosis. Twenty-one patient (91.3%) underwent surgery, of which 9 had incomplete resection.

Radiotherapy (RT) was delivered to 18 patients at a mean dose of 54 gy (30-54) with concomitant chemotherapy (CT) based on vincristine on 8 patients.

Twenty-one patients received adjuvant chemotherapy based mainly on VP16-carboplatin-cyclophosphamide and vincristine (52.3%).

Ten patients have relapsed after a median delay of 6 months (1-26), two patients were operated, four patients had CT, and none received RT.

After a mean follow up of 37 months, twelve were in complete remission,9 deaths were noted, and 2 patients were lost of follow up.

The 5-year-overall survival and the 5-year relapse free survival were respectively 48% and 44%.

**Conclusion:** A multimodal treatment have improved survival and prognosis. In our study, the recurrence rate seems to be higher and the overall survivor seems to be poorer which can be related to incomplete surgery. Broader studies are needed to more describe and understand these tumors.

## **Abstract 45**

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**Title:** Optic pathway gliomas in children: about 3 cases

**Theme:** Pediatric Neuro Oncology

**Title:** "Optic pathway gliomas in children: about 3 cases."

**Introduction:** Optic pathway glioma is a rare pathology in children with only a few cases described in infants. By way of three patients treated in the radiotherapy department of CHU Farhat Hached Sousse, we discuss their clinical and radiological characteristics, their treatment and course.

**Methods:** We report two cases of optic nerve gliomas and one case of chiasm glioma. The first case is about a 4-year-old girl followed since the age of 6 months for exophthalmos and bilateral nystagmus. The second is a 15-year-old girl followed for right divergent strabismus with right monocular blindness. The diagnosis was confirmed by MRI and histological findings in both cases. In the 3rd case, a 9-year-old boy presenting since the age of 2 years right palpebral ptosis with a progressive decrease in visual acuity. The diagnosis of an enormous glioma of the chiasma was confirmed by MRI, leading to reveal neurofibromatosis type 1.

**Results:** The first case was treated with exclusive radiotherapy. The second patient was referred by our service to ORSAY centre, where he was treated by proton therapy. The 3rd was admitted in a coma with bilateral mydriasis. He had an emergency ventriculoperitoneal shunt and then adjuvant radiotherapy. The radiation therapy dose was 54 Gy for all patients. The short-term outcome was favourable with lesion stability, despite a progressive recovery for the three children after one year.

**Conclusion:** The diagnosis and management of this rare tumor remain complex and controversial. Since the prognosis is poor, analysis of more cases is needed to identify better practices in the management of this tumor and to underline the place of radiotherapy.

**key words:** glioma, optic pathway, radiotherapy.



## Abstract 46

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**Title:** Radiotherapy for pediatric ependymoma

**Theme:** Pediatric Neuro Oncology

**"Introduction:** Ependymoma is the 3rd most common central nervous system tumor, and half of these are below 5 years of age. The prognosis of ependymoma remains poor. Factors associated with improved survival rate were essentially: total tumor removal and low histological grade. Surgical excision followed by radiation therapy was the primary mode of treatment. The place of chemotherapy remains to be defined.

**Purpose:** To describe epidemiological, clinical characteristics and therapeutic results of pediatric ependymoma in a Tunisian population.

**Methods:** Between 1995 and 2018, 8 pediatric ependymomas treated in the Department of radiation oncology farhat hached hospital, sousse, tunisia were retrospectively reviewed.

**Results:** There was 5 males and 3 females with a ratio of 1.66. The average age at diagnosis was 6 years (range, 1-14 years). An infratentorial origin was identified in most cases (62.5%). 4 patients had anaplastic tumor. Gross total resections were performed in all patients. 3 patients died before starting radiotherapy. 3 patients had post operative radiotherapy and 2 patients received adjuvant chemotherapy and post operative radiotherapy. Patients received focal radiotherapy on the bed tumor with doses between 54Gy and 59.4Gy using two-dimensional radiotherapy (2D-RT) for 2 patients and three-dimensional conformal radiotherapy (3D-CRT) for 3 patients. Spinal irradiation was performed according to various prognostic features in 2 cases. Acute complication was mostly transient and tolerable. No major late radiation effect was found. After a Median follow-up of 57.6 months, 4 patients were in complete remission and 1 patient died.

**Conclusion:** In our patient population, the characteristics of pediatric ependymoma are generally similar to those reported in the literature, with only minor differences. The prognosis remains poor with 3 – 7 years disease free survival, DFS has been reported to be ranging from 30% to 61%.

**Keywords:** Pediatric ependymoma; Radiotherapy; prognosis.

## Abstract 47

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**Title:** “Radiotherapy for Pediatric craniopharyngioma: About two cases.”

**Theme:** Pediatric Neuro Oncology

**"Introduction:** Despite its benign histopathology, Craniopharyngioma is an aggressive affection due to its location, its high recurrence rate, and long-term toxicity of combined limited surgery and irradiation or radical surgery. The prognosis is beneficial with low rate of mortality (3 to 12%).

**Methods:** Between 1996 and 2017, two craniopharyngioma cases were treated in the pediatric oncological radiotherapy department, Farhat Hached Hospital, Sousse Tunisia.

**Case1:** a thirteen-year-old girl had sellar and suprasellar craniopharyngioma treated with surgical resection. Three years later, she had headaches, raised intracranial pressure and vision symptom. She was referred to our department for second recurrence of craniopharyngioma in the MRI (Magnetic resonance imaging) finding, distorting the optic chiasma the third ventricle and the internal carotid artery. She underwent an uncompleted tumor resection because its location next to the carotid artery. She received tumor bed irradiation at the dose of 60 Gy in 30 fractions. After a median follow up of 229 months, she is in a complete remission.

**Case 2:** a twelve-year-old girl had headache and decreased visual acuity, the MRI revealed intra and suprasellar process in favour of craniopharyngioma which was treated with limited excision complicated by cecity. Six years later, she had intracranial hypertension syndrome. She received intratumorally cure of chemotherapy(bleomycin) with partial regression. For her third recurrence revealed on MRI with an obstructive hydrocephalus, she received radiation of the tumor at the dose of 52,2 Gy delivered on 29 fractions. After a median follow up of 30 months, she is in complete remission.

**Conclusion:** Primary surgery followed by radiotherapy is a powerful strategy for the treatment of craniopharyngioma, so, the management of this affection must be provided by a multidisciplinary team.

**Key words:** pediatric craniopharyngioma, surgery, radiotherapy.

## Abstract 48

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**Title:** “Ganglioneuroblastoma of the brainstem: A case report.”

**Theme:** Pediatric Neuro Oncology

**keys words:** Ganglioneuroblastoma,Brainstem, Radiotherapy.

**Introduction:** Neuroblastic tumors are classified into 3 histological groups: neuroblastoma, ganglioneuroma and ganglioneuroblastoma. The localization of a ganglioneuroblastoma in the brainstem is exceptional. We report a case of ganglioneuroblastoma of the brainstem in a 3-year-old girl.

**Observation:** A 3 year-old-girl, without notable pathological history, presented in July 2019 for diverging strabismus on the left eye. The neurological examination revealed an oculomotor paralysis. A cerebral CT scan returned without abnormalities. Additional MRI showed an extra-axial lesion developing at the level of the lateralized inter-peduncular cistern, suggesting a left III neuroma.

The tumor was inaccessible, inoperable, benign in appearance and slowly growing. The course was marked by the onset in November 2019 of left ptosis, right hemi body tremor, falling objects in the right hand and heaviness in the right lower limb. A second MRI revealed an extra-axial lesion of the left ponto-cerebellar angle which is progressing in terms of size and extension. A stereotaxic brain biopsy was taken with an anatomopathological and IHC examination in favour of a brainstem ganglioneuroblastoma. the patient underwent craniospinal axis radiotherapy (30 Gy) concomitantly with CT with good tolerance and clinical improvement. She received 5 courses of chemotherapy including, Vincristine, Etoposide, cyclophosphamide and carboplatin with severe side effects.

In addition, the present follow-up MRI showed a stability in size of the tumor.

**Conclusion:** Ganglioneuroblastoma is a rare type of primary neuroectodermal tumor (PNET) affecting almost exclusively infants and young children. The localization in the brainstem is exceptional and of poor prognosis because of the vital risk which it generates, the inaccessibility to the surgery and the drug resistance.

## Abstract 49

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**Abstract Title:** “Primary CNS Burkitt's Lymphoma in Children: A Rare Entity.”

**Abstract Theme:** Pediatric Neuro Oncology

**Background:** Primary central nervous system (CNS) lymphoma (PCNSL), an uncommon variant of extra-nodal non-Hodgkin lymphoma (NHL), can affect any part of the neuroaxis including the eyes, brain, leptomeninges, or spinal cord. It accounts for approximately 3% of all the primary CNS tumors diagnosed each year in the United States. Owing to the rarity of PCNSL, the disease has been challenging to study and an effective standard of care has been difficult to establish. Unfortunately, although durable remissions may be achieved for some patients with PCNSL, the tumor relapses in most cases. 1 Given the rare occurrence of this tumor, there are no prospective clinical trials in pediatrics to guide management. There have been case reports and case series describing approximately 100 cases over the course of last 20 years.

**Case Report:** A 6-year-old boy with no significant can't past medical history presents with headaches and vomiting, loss of walking and urinary retention and is found to have a 2- 3-cm left frontal lobe mass with spinal metastatic disease. A spinal mass biopsy reveals Burkitt's lymphoma (CNS BL). Cerebrospinal fluid cytology, as well as bone marrow biopsies are negative, and a whole-body MRI and Pan-CT scan does not demonstrate other areas of disease.

**CASE History and Course of Illness:** A 6-years-old boy presented September 2019 with history of intermittent headache and vomiting for past two weeks. These episodes were partially resolved on taking different medicines from local doctors. Later on, he developed backache increasing in intensity gradually accompanied by lower limb weakness and loss of walking and urinary retention.

He was further investigated for lower backache and loss of walking in the maintenance therapy for suspected diagnosis of ADEM by primary treating doctor. Eventually spinal biopsy was done three months later as he got deteriorated clinically and parents consented for biopsy. The patient was started on chemotherapy using UKCCSG Burkitt's Lymphoma protocol Group C with high dose methotrexate and improved and responded well on chemotherapy.

**Conclusion:** Though CNS primary lymphomas are very rare, but a high threshold of suspicion should be kept by the paediatricians and paediatric neuro-oncology teams to diagnose and treat them timely and efficiently in resource-limited settings.