CORE
Metadata, citation and similar papers at core.ac.uk
Provided by Elsevier - Publisher Connector

protocols used: EuroEwing-99 (10;43%), MSKCC-P6 (9;39%), others (4;17%).
Multimodal treatment was used in 23 (85%): chemotherapy and radiation in 8 (34%), chemotherapy and surgery in 5 (21%), chemotherapy, surgery and radiation in 10 (43%). Nine patients with non-axial disease and 5 patients with axial disease had surgery (p=0.03). Median follow-up for the treated patients was 27.5-months (Range:1-90).Out of 23 children who received curative treatment-11(47%) were alive & well, 5 (21%) were on treatment, 2 (0.08%) abandoned treatment & 4 (0.17%) had relapse/refractory disease; one (0.04%) patient died from treatment related toxicity. Five-year overall survival and event-free survival were 40% & 52%, respectively. Axial location (p=0.27), metastatic disease (p=0.74), and lack of surgery (p=0.34) were not statistically predictive of relapse/progression, probably due to low numbers of patient and limited duration of follow up.
Conclusions: In spite of a similar clinical profile and disease stage, the outcome of Ewings sarcoma in our patients simulates that reported in Indian studies, however lags behind Western statistics. Reducing abandonment by tracking defaulters and providing support, using a uniform protocol, and improving supportive care, were identified as plausible steps to improve outcomes.

ST-1_V1.2
CLINICAL EXPERIENCE AND OUTCOME OF HEPATOBLASTOMA IN CHILDREN: A 9-YEAR RETROSPECTIVE STUDY

Sanjeev Khera, Amita Trehan, K.L.N. Rao, Deepak Bansal, Nandita Kakkar, Radhika Srinivasan. *PGIMER, Chandigarh, India*

Background: Hepatoblastoma (HBL) is the most common primary liver tumor in children. Metastatic disease at presentation, alphafetoprotein (AFP) less than 100 ng/ml, PRETEXT IV, undifferentiated histology are usual poor prognostic markers. Neoadjuvant chemotherapy with surgical resection has led to increased survival of these patients
Aim: This single centre study assesses the outcome and the factors affecting prognos is of children treated as per guidelines of SIOPEL3 in a low middle income country (LMIC).
Materials & Methods: Forty children with HBL treated as per protocol from Jan 2007 to Dec 2015 were analyzed. The diagnosis was established by imaging, AFP and histology/cytology.
Results: 27 boys & 13 girls; median age 12 months (2-120) with a mean symptom diagnosis interval (SDI) of 5.3 weeks (1-24) were diagnosed. Multifocal involvement was seen in 7/40. Median AFP at diagnosis: 16500 ng/ml (3-406198). 2 had AFP less than 100 ng/ml. Mean platelet count at diagnosis was 701375/cmm (95% CI 595510.80 to 807239.20).7 (17.5%) had metastatic disease (6 lungs; 1 adrenal). PRETEXT: I-IV: 6, 13, 11 & 9. Nineteen: high risk (HR) disease (7 with SDI more than 8 weeks) 13/40 defaulted/refused therapy. All cases received neoadjuvant chemotherapy. 3 deaths occurred prior to surgery. 25 underwent surgery; complete resection (CR): 18/25 [14 : well; 3 relapsed, 1 died]. 7 cases subtotal resection : 2 : well, 4 had persistant disease and 1 death.2/3 children who received salvage chemotherapy with Docetaxel had no response. HR patients had a poorer outcome (p=.012), more multifocal disease (p=.0395) and significantly lower CR (p=.0016) as compared to standard risk patients. On comparing well Vs relapsed/residual disease cases; there was no difference in AFP levels, volume reduction after neoadjuvant therapy, metastasis, & histopathology (p=ns). Relapsed/residual disease was significantly higher in those with higher mean age (24 mth Vs 12 mth); PRETEXT IV & multifocal disease (p=0.02 & 0.04). 16 of 27 patients are presently well; median follow up being 48.3 months (7-101).
Conclusion: 59 % (16/27) of cases who were treated are well. 86%of SR HBL had good outcome. Higher age at presentation, multifocality and high PRETEXT was associated with poor outcome. HR patients had a longer SDI. Docetaxel as a salvage therapy was not effective. Liver transplant, not available at our centre, would have helped the children with incomplete resection. Delayed referral and diagnosis remains a problem in LMIC's as evident by greater SDI in those with HR disease.

ST-1_V1.3
MULTIDISCIPLINARY MANAGEMENT OF HEPATOBLASTOMA WITH INCORPORATION OF LIVER TRANSPLANTATION IN CHILDREN

Vimal Kumar[2], Naresh Shanmugam[1], Julius Xavier Scott[2], Mukul Vij[3], Priya Ramachandran[4], Gomathy Narasimhan[4], Mettu Srinivas Reddy[4], Venugopal[4], Deenadayalan Munirathnam[3], Chayarani Kelgeri[1], Karthick Sundaram[1], Mohamed Rela[1]. [1] *Department of Pediatric Gastroenterology, Hepatology and Nutrition, Global Health City, Chennai, India;* [2] *Department of Pediatric Hematology and Oncology, Global Health City, Chennai, India;* [3] *Department of Histopathology, Global Health City, Chennai, India;* [4] *Institute of Liver diseases & transplantation, Global Health City, Chennai, India*

Abstract
Background: Advances in chemotherapy, liver resection techniques and pediatric liver transplantation have vastly improved survival in children with hepatoblastoma (HB). These are best managed by a multidisciplinary team in a setting where all treatment options are available. Until recently this was difficult to achieve in India.
Methods: Review of all children (< 16yrs) with HB treated in a pediatric liver surgery & transplantation unit between Jan 2011 and July 2016. Data regarding the clinical presentation, pre-operative management, surgical treatment, postoperative course and outcomes was extracted from a prospectively managed database.
Results: Thirty children were treated for HB during the study period. Nine children were PRETEXT 4, 7 were PRETEXT 3, 13 were PRETEXT 2 and one was PRETEXT 1. All children received neoadjuvant chemotherapy before surgery followed by adjuvant chemotherapy. Nineteen children had complete resection, while six underwent primary living donor liver transplantation. There were 6 mortalities including 5 children who poorly responded to chemotherapy with progressive tumor extension. At a median follow-up of 30 months, two children who underwent resection and one child who underwent liver transplant had disease recurrence.
Conclusion: Improved outcomes can be achieved in children with HB even in countries with limited resources when they are managed by multidisciplinary teams with expertise in pediatric oncology, liver resection and liver transplantation.

ST-1_V1.4
CLINICOPATHOLOGICAL PROFILE OF PEDIATRIC MEDULLOBLASTOMA

Rikki R. John[1], Leenu Joseph[1], Ari G. Chacko[2], Ranjit Moorthy[2], Magesh P[3], Geeta Chacko[3], Leni G. Mathew[1]. [1] *Pediatric Hematology/Oncology, Christian Medical College, Vellore, India;* [2] *Neurosurgery, Christian Medical College, Vellore, India;* [3] *Neuropathology, Christian Medical College, Vellore, India*

Introduction: Medulloblastoma is the second most common primary malignant brain tumour in children. We present the clinic-pathological profile of children seen in a pediatric hematology/Oncology unit over a ten-year period.
Method: Clinical and laboratory data of children with medulloblastoma who attended the pediatric hematology/oncology unit of CMC, Vellore from 2004 to 2014 were retrieved from the medical records and reviewed.
Results: 76 children, 49 boys and 27 girls, with mean age at presentation 8.5±3.45 years. 7 were < 3 years and 22 were >10years. Headache (86%) vomiting (79%) and ataxia (58%) were the common symptoms. The mean duration from onset of symptoms to diagnosis was 2.7 ±2.2 months with 78% diagnosed within 3 months. All children presented to the neurosurgical team at first visit. 30% required emergency CSF diversion procedure. All children underwent upfront surgical excision; 30 had residual tumour >1.5cm^2. Risk stratification based on age at presentation, volume of residual tumour and presence of metastatic disease, showed 43/76 to be in the high risk category.
Histological classification showed the following: classic 22 (37%), desmoplastic 19 (33%), medulloblastoma with extensive nodularity9 (16%), large