cell 5 (9%) and anaplastic 2 (4%). 19 cases were only reported as meduloblastoma WHO IV. Using immunohistochemical marker beta catenin and histology 43 tumours were subdivided into WNT (26), SHH (13), non-WNTnonSHH (4) groups.

**Conclusion**: There was an obvious male predominance in this group. 78% of cases were diagnosed within 3 months of onset of symptoms. 57% of cases had high risk disease.

### ST-1\_V1.5

### INFLAMMATORY MYOFIBROBLASTIC TUMORS: A TALE OF THREE STORIES

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**Background/Objectives**: Inflammatory myofibroblastic tumors (IMT) are unusual intermediate malignant potential tumors that occur in soft tissues and visceral organs of children and young adults. The tumors consist of myofibroblastic spindle cells and inflammatory cells. These tumors can be locally aggressive, recur or rarely metastasize. There are no clear consensus guidelines on treatment for patients with such tumors. We describe three cases from our tertiary academic paediatric institute in Singapore.

**Design/Methods**: A retrospective analysis of paper, electronic records, and histology slides of patients diagnosed with IMT in our hospital between 1997-2012 was performed.

Results: Three patients with IMT were identified. All patients had intraabdominal tumors (omentum, spleen, supra-renal) with one patient also having a concurrent IMT located in the left hemi thorax. Histopathological findings reported positivity for ALK -1 stain in 2 patients. Primary complete surgical resection of the tumors was attempted without success. And hence adjuvant chemotherapy was administered in all cases. A variety of chemotherapeutic agents in combinations were used as per physician preference. Two patients also received monotherapy with nonsteroidal anti-inflammatory drug (NSAID) when the tumors did not respond to chemotherapy. Both the patients on NSAID therapy showed transient partial responses. One patient with aggressive IMT died due to neutropenic sepsis. The mTOR inhibitor sirolimus was started on one patient who is now alive with stable disease three years after commencement. A local recurrence in one patient was treated with further surgery and she now remains in complete remission. These two patients remain on active follow up.

**Conclusion**: Sirolimus appears to offer disease control in children with inoperable or resistant IMT. Further studies are required to understand and manage these fascinating mesenchymal tumors better.

### ST-1\_V1.6

## OUTCOMES OF SURGERY FOR RENAL TUMOURS WITH INTRAVASCULAR EXTENSION

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**Aim**: The aim of this study was to review the management of children with Wilms' tumour who had intravascular thrombus.

**Material and methods**: The data regarding presentation, treatment received, surgical management, complications and outcomes were evaluated. All patients received neoadjuvant chemotherapy except in two.

**Results**: The study cohort included 31 patients with intravascular thrombus treated from 2006 to 2015. The thrombus extent at presentation was: Infrahepatic inferior vena cava (IVC) -19, retrohepatic IVC -6, suprahepatic IVC -1 and Atrium -5. There was complete clearance of IVC in 11/31 patients with neoadjuvant chemotherapy. Regression of the thrombus occurred in 17/31 patients. Due to regression of the tumor thrombus from the atrium, cardiopulmonary bypass could be avoided in 2 patients. In all patients the thrombus had to be dissected off from the tunica intima due to

dense fibrosis around it. The only major complication was massive bleeding in one patient with atrial thrombus. There was no perioperative or 30-day postoperative mortality. The 3-year OS and EFS was 89.3% and 77.8% respectively.

**Conclusions**: Intravascular tumor thrombus extension has favourable outcomes after contemporary multidisciplinary treatment. Chemotherapy aids in surgery, with tumor regression and thus may obviate the need of cardiopulmonary bypass in atrial thrombus.

### ST-1\_V1.7

# PEDIATRIC MEDULLOBLASTOMA: EXPERIENCE AT TATA MEDICAL CENTER, KOLKATA

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**Background**: Medulloblastoma is the commonest pediatric malignant brain tumor. Cure-rates of ~75% have been achieved in the West. Indian centers have reported inferior survival. The aim of the study was to analyze the clinical profile and outcome in children treated at a new referral cancer hospital in Eastern India.

**Methods**: A retrospective analysis of case records of children (<18-years) diagnosed with medulloblastoma, between June 2011-July 2016, was performed. Modified Chang staging, using MRI of brain and spine, and analysis of cerebrospinal fluid was used. Children aged >3-years, with post-operative residual tumor <1.5cm², M0 stage, and non-large cell/anaplastic histology, were classified as 'standard-risk.' 'Standard-risk' and 'high-risk' children received cranio-spinal irradiation with 23.4Gy in 13 fractions, and 36Gy in 20 fractions, over 3.5-4 weeks, respectively. Posterior fossa and/or tumor bed was boosted to a total dose of 55.8Gy in 31 fractions. Following radiation, 'standard-risk' children received 8 cycles of cisplatin, lomustine and vincristine. 'High-risk' patients received 8 cycles of cisplatin, cyclophosphamide and vincristine, or, ifosfamide, cisplatin and etoposide (physician's discretion). Analysis was performed using IBM-SPSSv20. Kaplan-Meier method was used for survival analysis.

**Results**: Twenty-six children were enrolled. Median age was 6-years (range:0.9-13.5); 4 (15.3%) were  $\leq$ 3-years at diagnosis. 54% were males. Median symptom-interval was 3.7-months (range: 1-12). Symptoms included vomiting (20; 77%), headache (17; 65%), unsteadiness (14; 54%) and cranial nerve palsy (2; 7%). Location of tumor (n=22) included midline, cerebellum (9; 41%), lateral cerebella (5; 23%) and roof of 4<sup>th</sup> ventricle (8; 36%). Modified Chang staging: T (n=17): T1 (1; 6%), T2 (2; 12%), T3 (5; 29%), T4 (9; 53%), and, M (n=22): M0 (9; 41%), M1 (3; 13.5%), M3 (7; 32%), M4 (3; 13.5%).

Seven (32%) children were standard-risk and 15 (68%) were high-risk; data was missing for the rest. Eight (31%) refused treatment. Extent of surgical resection (n=16) included gross total resection: 7 (44%), near total resection: 3 (18.5%), sub-total resection: 6 (37.5%). A ventriculo-peritoneal shunt had been inserted in 11/20 (55%). Median time from surgery to initiation of radiotherapy was 41-days (range:34-51).

At a median follow-up of 28.5-months (range:0.1-55), 12 (46.5%) were alive, 9 (35%) of whom had completed, and 3 (11.5%) were on treatment; 3 (11.5%) had relapse/progression, 2 (7.5%) had abandoned treatment, while 1 (3.5%) had received palliative care. Among the 4 children  $\leq$ 3-years, 1 had disease progression and 3 refused treatment. 4-year EFS (for those who completed therapy was  $66.1 \pm 1.2\%$ ; standard-risk: 100%, high-risk:  $45 \pm 1.8\%$ . Treatment-related complications included posterior fossa syndrome (7; 39%), sepsis (4; 22%), shunt obstruction (1; 5%) and hearing loss (4; 22%).

**Conclusion:** Limitations of the study included high prevalence of treatment-refusal/abandonment (10; 38%) and non-availability of molecular subtyping for more accurate risk stratification. Nevertheless, this study shows that when properly diagnosed and treated, the outcomes of children with standard risk medulloblastoma in India are comparable with the west. Further improvements in outcome require providing social support to reduce treatment-refusal/abandonment, standardization of care for

high-risk disease, reduction of time from surgery to radiotherapy, and long-term follow-up for late effects.

## Transfusion Medicine & Supportive Care TM\_SC-1\_V1.1

CLINICAL EXPERIENCE OF GRANULOCYTE TRANSFUSION THERAPY IN MANAGEMENT OF NEUTROPENIA RELATED INFECTIONS IN A TERTIARY CARE CENTER

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**Background**: Granulocyte transfusions have recently gained popularity for the treatment of severe infections in neutropenic patients which do not respond to conventional antimicrobial therapies and recombinant growth factors. We conducted the present study to determine the clinical course of patients with neutropenia related infection receiving granulocyte transfusions in our center and to assess its efficacy in resolution of infections.

**Methods**: Retrospective analysis of all pediatric patients up to 18 years of age with neutropenia and infection receiving granulocyte transfusions in our hospital between January 2015 and July 2016.

Results: Twenty one pediatric patients with severe neutropenia related infections unresponsive to appropriate antimicrobial agents were included in the study. They received a total of 57 granulocyte transfusions. The study population comprised of 10 females (47.6%) and 11 males (52.4%). Median age of the study population was 9 yrs (5 months-16 years). 13 patients suffering from nonmalignant hematological diseases and 8 patients with malignant diseases were included in the study population. Voluntary healthy donors were used after granulocyte mobilization using granulocyte colony- stimulating factor (G-CSF) and dexamethasone. The median donor WBC count before leukapheresis was  $31.9 \times 10^9$  /L and the mean donor granulocyte yield was  $8.9 \times 10^{10}$  /L. Seven patients had localized infection while 14 had sepsis. Causative organisms could be isolated in 16 cases out of which gram negative bacillus was isolated in 15 (93.75%). Median duration of neutropenia, antimicrobial therapy and G-CSF administration before granulocyte transfusion was 11 days (range 2-34), 10 days (range 3-20) and 9 days (range 2-20) respectively. Patients received a mean of 2.85 (range 1-8) granulocyte transfusions and mean cell dose of  $2.7 \times 10^{10}$  granulocytes. Ten of the total twenty one patients included had a favorable response and recovered from the infection. Granulocyte transfusions were generally well tolerated in most of the cases except for one episode of transfusion associated acute lung injury.

**Conclusion**: Granulocyte transfusions seem to be a clinically useful and generally safe adjunct in management of severe neutropenia related infections.

#### TM\_SC-1\_V1.2

BLOOD AND BLOOD PRODUCTS TRANSFUSION AUDIT: CURRENT PRACTICES AND LACUNAE IN BLOOD AND BLOOD PRODUCT TRANSFUSION AMONG HOSPITALIZED CHILDREN

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Transfusion audits are most effective tools to detect inappropriate blood product transfusion practices and suggest remedial measures to ensure safe and effective use of this precious product in health-care.

**Objectives**: Present study was designed to assess current practices regarding blood product transfusions in children with special emphasis on the appropriateness of these transfusions.

**Materials & Method**: Present study was a prospective study conducted over a period of 22 months from January 2013 to October 2014. Each episode of transfusion was analyzed and divided into appropriate and inappropriate according to the type of blood component, the clinical and hematological indication. Data was reviewed according to British Committee for Standards in Hematology and American Association of Blood Bank guidelines.

**Results**: In this study, a total 741 transfusions were used in 327 cases, including 449 packed red blood cells (60.6%), 148 Fresh frozen plasma (20.0%), 140 (18.9%) platelets and 4 (0.5%) whole blood transfusions. Appropriate usage of blood and blood products was 70.5%. Most inappropriately used was FFP (39.2% of FFP transfusions), followed by Packed RBCs (28.7%) and Platelets (22.9%). Packed RBCs transfusions were used most commonly for sepsis with/without DIC (25.2%), followed by nutritional anemia (20.7%) Packed RBC transfusions were used most inappropriately for nutritional anemia (49.5%) and sepsis (46.9%). FFP transfusions were used maximally for the indication of sepsis with/without DIC (43.2%), followed by perioperative indications (21.6%). FFP transfusions were most inappropriate for Dengue (68.8%) and perioperative indications (53.1%). Platelet transfusions were most commonly used for Aplastic anemia (45%) followed by sepsis (24.3%) Platelet transfusions were most inappropriate in Dengue cases (85%) and Idiopathic thrombocytopenic purpura (50%).

**Conclusion**: Present study concludes that in the study population, about one-third of all blood component transfusions used in study center are inappropriate. This study suggests establishment of set protocols for requisition of blood component transfusions in the study set-up to minimize irrational and unsafe use of these products.

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#### TM SC-1 V1.3

# FUNGAL BRAIN ABSCESS IN HAEMATOLOGICAL MALIGNANCY: IS GOOD OUTCOME POSSIBLE?

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**Introduction**: Incidence of invasive fungal infections in children with hematological malignancies is increasing. We present clinical features and outcome of children with fungal brain abscess in acute lymphoblastic leukemia (ALL), an entity which traditionally has dismal outcomes.

**Methods**: Eighteen months data is presented. Total 160 new ALL patients were recruited. Thirteen developed proven fungal infection. Details of children with fungal CNS infection were retrieved and analyzed.

Results: Five children had fungal brain abscesses. B-cell ALL:T-cell was 4:1. Three children were standard risk, 2 intermediate risk. All were on intense chemotherapy including steroids; 4 induction, 1 intensification. Clinical presentation: Prolonged neutropenia: 100%; fever: 100%; altered sensorium/seizures seen in 80% (4/5). Duration of neutropenia: 16 days (14-23). Primary focus was lung in 4, ear in 1. Brain abscess was single in 2, multiple in 3 children. Meningitis was present in 2. All children underwent surgery (burr-hole drainage:4, excision:1). Culture/PCR proven fungus was seen in all: Aspergillus in 4; Mucor:1. Elevated serum galactomannan seen in 66%. Combination antifungals were given for aspergillus: Amptotericin (plain/liposomal)& voriconazole for 10 days, followed by voriconazole alone for 6 months. Single mucor patient received amphotericin for 12 months. Overall survival at 10 months: 80%; event free survival: 40% (eventsdeath:1, defaulter:1, cortical blindness:1). Three children are continuing chemotherapy.

**Conclusion**: Mortality rate in fungal brain infections in children is reported to be 65%. Better outcomes in our patients can be attributed to multimodality therapy and early initiation of antifungal therapy along with judicious management of chemotherapy.

**Keywords**: Aspergillus, acute lymphoblastic leukemia, brain abscess, invasive aspergillosis