Variance (ANOVA) for comparison of means across more than two groups. The proportions were compared using the chi square test or the Fisher's exact test (for low expected cell counts).
Results: The mean age of the study group was 8.08 years, while that of the control group was 8.88 years. Eighty percent had Hemophilia A and 20% had Hemophilia B. Severe hemophilia was seen in 87% and 13% had moderate Hemophilia. The mean scores of the child self-report and parent proxy report (results of the questionnaire answered by the parent) of the study group was 79.81 and 72.75 respectively, which was significantly lower as compared to the control group, suggestive of impaired quality of life .The mean scores of the parent proxy report were significantly lower than the child report scores for both the study and control groups, indicating parents reporting of impaired quality of life of the affected children. Joint bleeds were present in 36.66%, 73.3% had muscle bleeds and 16.66% had intracranial bleeds. Target joints were present in 13.33%. The median number of hospitalizations was 3 from date of diagnosis till the inclusion of the patients in the study group. The quality of life was significantly affected in children with more number of bleeding episodes and with the presence of target joints. The qol was not significantly affected by the age of diagnosis, duration of illness and the socioeconomic status.
Conclusion: The Quality of Life of children with Hemophilia is significantly impaired as compared to the general population. Number of Hospitalizations, presence of target joints, number of bleeding episodes, age at diagnosis and duration of disease were associated with the quality of life of children with hemophilia and hence rehabilitation measures should be encouraged along with primary factor prophylaxis and encouraging the patients to learn home treatment to improve scores and quality of life.

BDT-1_V1.3
GLANZMANN'S THROMBASTHENIA – CLINICAL PROFILE OF PATIENTS IN A TERTIARY CARE CENTRE IN SOUTH INDIA

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Abstract
Objectives: To describe the clinical profile of Glanzmann's Thrombasthenia (GT) in a tertiary care centre in South India.
Methods: Retrospective descriptive study done by chart review of all patients on follow up between Jan 2005 – Aug 2016. Study population included all children less than 18 years of age diagnosed at our centre. A diagnosis of GT was made based on standard criteria.
Results: A total of 46 patients (representing 42 families) are being described in this study. Mean age at presentation was 4 years. 65.2% (30/46) had first bleed within the first two years of life. The male to female ratio was 1.4:1. 21/46 children (46%) were born out of consanguineous marriage. The common symptoms were epistaxis, gingival bleeding and skin bleeds. Gastrointestinal bleeding in the form of hematemesis and melena was frequent, hemarthrosis and intracranial bleed was rarely seen while muscle involvement and hemoptysis was not seen. Post surgical bleed in the form of tonsillar bleed following tonsillectomy was common. Presentation in the neonatal period was rare (<1%) and was observed in the form of purpura, epistaxis and intracranial bleed. Menorrhagia was the most common presentation in children first presenting in teenage. 25% had life threatening bleeds while 50% had growth retardation due to chronic anemia.
Conclusion: Knowledge of the clinical profile of this disease will aid in suspecting and promptly evaluating the disease as early diagnosis aids in preventing life threatening bleeds.

BDT-1_V1.4
RARE PRESENTATION OF A RARE BLEEDING DISORDER

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Background: Factor V deficiency (Parahemophilia) is a rare autosomal recessive bleeding disorder with an incidence of 1 in million persons. The most common bleeding symptoms are mucocutaneous bleeds, menorrhagia and post surgical bleeds. There are anecdotal case reports of life threatening bleeds in severe Factor V deficiency. We present two young infants with prolonged PT/aPTT with this rare bleeding disorder causing Intra cranial hemorrhage.
Methods: We report two children with severe Factor V deficiency with severe unusual bleed presented to St John's Medical College Hospital, Bengaluru.
Case 1: A 3 months old baby boy with uneventful perinatal period presented with complaints of seizures. He was in altered sensorium with severe pallor, bulging anterior fontanel, unequal pupils and exaggerated DTR. Initial blood investigations revealed severe anemia, prolonged PT (70 sec), aPTT (120 sec), INR (9) and normal TT (13.7 sec). Neuroimaging (CT Brain) showed left intra-cerebral bleed extending from the frontal lobe to the parieto-occipital region with 5mm shift of the midline to the right. A possibility of Late onset VKDB or Common pathway deficiency was considered in view of no improvement in PT/aPTT with Inj Vit K and FFP. Factor assay were sent which showed Factor V levels of <1%.
Case 2: A 4 day old baby girl was referred to our institute with complaints of bleeding from nose and umbilical stump. Child had received Inj Vit K at birth. On examination the child was active, alert with few ecchymotic patches over the limbs and cephalhaematoma. Initial blood tests revealed anemia, prolonged PT (30 sec), aPTT (78 sec), normal TT. Child was managed with PRBC transfusion, Inj Vit K and other supportive measures. On the same day she developed massive intra cranial hemorrhage leading uncal herniation and succumbed.
Conclusion: Considering bleeding disorder other than VKDB in neonates and young infants with prolonged PT/aPTT is important in diagnosis of rare bleeding disorder with life threatening complications.

BDT-1_V1.5
PROFILE OF THROMBOCYTOPENIA IN INPATIENT CHILDREN WITH ACUTE FEBRILE ILLNESS IN A TERTIARY CARE CENTRE

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Background: Acute febrile illness with thrombocytopenia is a common problem with increased mortality and morbidity if not diagnosed and treated promptly. There is a need to ascertain the prevalence and causes of febrile thrombocytopenia to enable monitoring and prompt treatment.
Objectives: To estimate the prevalence and identify the underlying etiology of thrombocytopenia in children presenting with acute febrile illness, and to determine the need for platelet transfusion.
Materials and Methods: 192 inpatient children in the age group of 3 months to 12 years presenting with fever of 1-14 days duration over a period of 3 months were included in the study. The discharge cards of the recruited subjects were reviewed. Platelet counts at admission and during hospital stay were noted. Platelet transfusions, when given, were documented. The etiological profile of the fever was assessed to evaluate the causes of thrombocytopenia. Chisquare test was used to test the association between thrombocytopenia and its different causes. p value <0.05 was taken as statistically significant.
Results: The hospital based prevalence of thrombocytopenia in acute febrile illness in our centre was 22.9% (n=192). The most common cause of thrombocytopenia was Dengue fever (38.6%) followed by Acute Febrile Illness of unknown etiology (29.5%). Other causes included Leptospirosis (6.8%), Malaria (9.2%), Pneumonia (9.2%), Bronchiolitis (4.5%) and UTI (2.2%). 9 out of 26 cases of Dengue fever were not associated with thrombocytopenia. On statistical analysis, there was a significant association between thrombocytopenia and Dengue fever ($p<0.01$) over the other causes of fever. The mean duration after onset of fever at which thrombocytopenia was detected was found to be Day 7. Platelet transfusion was required only in 1 patient in view of hematemesis and malena at a platelet count of 34,000/cu.mm.
Conclusions: Febrile thrombocytopenia is a common clinical condition requiring detection and monitoring. Infections, particularly Dengue, were