negative (LTCD3+ TCR αβ+ CD4- CD8-) about 27% (control < 2.5%). The study of FAS gene allowed the identification of a mutation in exon 9.

Conclusion ALPS is an underestimated entity that must be considered in non-malignant lymphoproliferation, autoimmunity and expansion of an unusual population of a/b CD3+/CD4-CD8- (double-negative T cells)>1%.

A RARE CASE OF LANGDON DOWN SYNDROME WITH COMPLETE ENDOCARDIAL CUSHION DEFECT, TETRALOGY OF FALLOT, DEFICIENCY OF FACTOR VII

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Aims We sought to summarize a very rare association between multiple rare incidence diseases in a patient with Langdon-Down syndrome and also to correctly document each pathology and use the best course of treatment.

Background Factor VII deficiency has an incidence of 1 in 500,000 reported cases. Complete endocardial cushion defect [ECD] occurs in 2% percent of all congenital heart defects. Additional cardiac abnormalities (persistent ductus arteriosus and tetralogy of Fallot [ToF]) may occur in 10% of all ECD’s. Associated defects are rare in children with Down syndrome.

Methods A 5 weeks old infant with a Down phenotype was admitted in the Intermediate Care Unit for severe tonic-clonic seizures and an unexplored heart murmur. A computed tomography scan revealed a massive hemorrhaging in the fronto-parieto-occipital left cerebral region. Trauma was excluded and the prothrombin time was prolonged with the activated partial thromboplastin time normal so we sent a blood sample for the factor VII activity.

We performed an echocardiography. A karyotype study was carried out.

Results

- Complete ECD with the common atrio-ventricular valve in dextroisomerism, left ventricle hypoplasia, associated with ToF
- The factor VII activity showed a 2% activity level
- Classical 21 trisomy

Conclusion We provided a good documentation of a very rare association between separate severe pathologies and we showed that when faced with a congenital malformative syndrome one should never stop looking for other abnormalities.

THALASSEMIA PREVENTION AND ACTIVITY OF PEDIATRIC HEMATOLOGY AND ONCOLOGY DEPARTMENT AT BANGBANDU SHEIKH MUJIB MEDICAL UNIVERSITY IN BANGLADESH

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Background and Aims Thalassemia is a genetic and crucial disease. Approximately 240 million people are suffering from this disease. Every year 10 million children are suffering from this disease. Hb-E disease is available in south east Asia, north east India and Bangladesh. Originally Hb-E disease is 5 times more than Beta Thalassemia in Bangladesh.

Method The outbreak of this disease is not calculated at this moment but carrier is 15 million. We are collected experimental data from 3 hundred volunteers in our center. They have not family history of Thalassemia. In that experimental data Beta Thalassemia carrier 2.83% and Hb-E Carrier 10%. If we are experiment among the people who have history of Thalassemia, this disease is increased no doubt. This disease have actually no curable treatment except Bone Marrow transplantation. Treatment cost is excessive and unbearable. Only time to time Blood transfusion and costly drug is given for the increasing of life span. Treatment cost of every 30kg child need 4 lac taka every year. If 2 bag blood need every patient in every month, 1 lac 20 thousand bag blood will be need every month in Bangladesh.