Mucopolysaccharidosis type I is caused by a deficiency of the lysosomal enzyme alpha-L-iduronidase. As a result, glycosaminoglycans progressively accumulate in the lysosomes and causes dysfunction of cell, tissue and organ. Glycosaminoglycan is a constituent of the extracellular matrix, connective tissue and joint fluid. Its’ progressive accumulation within the cells of various organs ultimately compromises their function. Mucopolysaccharidosis type I is caused by the deficiency of the lysosomal enzyme alpha-L-iduronidase. As a result, glycosaminoglycans progressively accumulate in the lysosomes and causes dysfunction of cell, tissue and organ.

Case report-
3 years old baby girl from Pakistan had fever since 1 year of age. Baby also had developmental delay, difficulty of vision and swelling in the back. Initially baby was managed symptomatically in Pakistan. In view of persistence of symptoms, she was brought to a tertiary care hospital in New Delhi for further management. On evaluation, she was found to have umbilical hernia, kyphosis, median nerve neuropathy and corneal clouding. Based on serum enzyme assay and excess glycosaminoglycan level in urine, she was diagnosed as a case of mucopolysaccharidosis type I. She was managed with few weeks course of enzyme replacement therapy and symptoms were persisted. After 5 weeks of last enzyme replacement, she was admitted to bone marrow transplant unit. After finishing the conditioning chemotherapy, allogenic unrelated bone marrow transplant was done. Immune suppressant therapy in form of injection Cyclosporin was started from day+9 of bone marrow transplant. But no improvement was found. Cyclosporin which was started two days before transplant was stopped and replaced by tacrolimus on day+1. But fever persisted. Immunological mechanism should be kept in mind. Increased immunosuppressant therapy will help to improve such group of patients.

DISCUSSION
Today, enzyme replacement therapy and haematopoietic stem cell transplantation are the standard options worldwide. Haematopoietic stem cell transplantation can treat the brain in the course of the disease, as stem cells can engraft and differentiate in the CNS. In contrast, infused enzyme replacement therapy can not cross the blood brain barrier easily due to large size. Following autologus transplant, patient may develop fever and skin rash. This may be due to engraftment syndrome. Release of proinflammatory cytokines, including interleukin-1 and tumor necrosis factor α are responsible for such happening. Our patient had fever, skin rash, diarrhea during neutrophil recovery period. But there was no such feature like pulmonary infiltrate, jaundice, weight gain, hypoalbuminaemia or neurological manifestation. Neutrophil recovered on Day+14 of post transplant. All the infective possible causes of fever were ruled out. Increased immunosuppressant therapy improved the symptom which was caused by non-infective immune reaction.

Conclusion-
Treatment of mucopolysaccharidosis type I disease is a challenge. Post stem cell transplantation, fever always may not be due to only infective cause. Immunological mechanism should be kept in mind. Increased immunosuppressant therapy will help to improve such group of patients.

REFERENCES