SO SHIVBALAN AND MV SRINATH
Sundaram Medical Foundation,
Dr Rangarajan Memorial Hospital,
Shanthi Colony, IV Avenue, Annanagar,
Chennai 600 040, Tamil Nadu, India.
sivabalan.somu@gmail.com

REFERENCES

1. Jadhav M., Webb JK, Vaishnava S, Baker SJ. Vitamin B12 deficiency in Indian infants. A clinical syndrome. Lancet. 1962;2:903-7.
2. Grasbeck R., Tanner SM. Juvenile selective vitamin B12 malabsorption: 50 years after its description – 10 years of genetic testing. Pediatr Res. 2011;70:222-8
3. Nyholm E, Turpin P, Swain D, Cunningham B, Daly S, Nightingale P, *et al*. Oral vitamin B12 can change our practice. Postgrad Med J. 2003;79:218-20.
4. Vidal-Alaball J, Butler CC, Cannings-John R, Goringe A, Hood K, McCaddon A, *et al*. Oral vitamin B12 versus intramuscular vitamin B12 for vitamin B12 deficiency. Cochrane Database Syst Rev CD004655 (2005). doi:10.1002/14651858.CD004655.pub2
5. Lin SH, Sourial NA, Lu KC, Hsueh EJ. Imerslund-Grasbeck syndrome in a Chinese family with distinct skin lesions refractory to vitamin B_{12}. J Clin Pathol. 1994;47:956-8.

Status of Primary Immunodeficiency Disorders in India

We read the recent article on approach to primary immunodeficiency disorders (PID) [1] with interest. These disorders are under-diagnosed in the developing world. On reviewing published Indian data on PID we came across only 4 large series reporting a total of 386 cases [3-5]. Most common PID's reported are disorders of immune dysregulation followed by phagocytic disorders and predominant antibody deficiencies [4,5]. High mortality rates of upto 51% have been reported [5].

We report here a series of 11 children (8 males) registered in our PID clinic from January to July 2013. Eight were males and three females. Mean age was 5.2 years (4 months-12 yrs.). Two had *X*-linked agammaglobuinemia. Both are on replacement intravenous immunoglobulin (IVIG) therapy and doing well. Third, a 12-year-old male presented to us with severe aplastic anemia (SAA) with common variable immunodeficiency. He underwent matched sibling donor bone marrow transplant (BMT) for SAA. At eighty days post-transplant, he has normal blood counts and immunoglobulin levels. Fourth child had pure red cell aplasia with isolated IgM deficiency. He responded well to prednisolone. Fifth child had congenital neutropenia and negative for ELANE mutation for congenital neutropenia. His neutrophils increased only after high dose granulocyte colony stimulating factor (60 ug/kg/day). Sixth patient had hemophagocytic lympho histiocytosis and was managed as per HLH 2004 protocol. Two infants were diagnosed as cases of autoimmune lymphoproliferative syndrome and are doing well on prednisolone and mycophenolate. Two were diagnosed as case of Heme-oxygenase-1 deficiency with auto-inflammatory syndrome. One boy was diagnosed with *X*-linked severe combined immunodeficiency and underwent matched unrelated donor BMT abroad without conditioning. Although his T-cells and NK-cells recovered but he still has low immunoglobulin levels.

Our small series shows that improvement in survival is possible; although, pan-India improvement would require increased awareness among pediatricians, establishing specific centers offering genetic diagnosis and definitive therapy like BMT.

YOGI RAJ CHOPRA AND SATYA PRAKASH YADAV
Pediatric Hematology and Bone Marrow Transplant Unit,
Fortis Memorial Research Institute, Gurgaon, India.
satya_1026@hotmail.com

REFERENCES

1. Madkaikar M, Mishra A, Ghosh K. Diagnostic approach to primary immunodeficiency disorders. Indian Pediatrics. 2013; 50: 579-586.
2. Verma S, Sharma PK, Sivanandan S, Rana N, Saini S, Lodha R, *et al*. Spectrum of primary immune deficiency at a tertiary care hospital. Indian J Pediatr. 2008; 75:143-148.
3. Gupta S, Madkaikar M, Singh S, Sehgal S. Primary immunodeficiencies in India: a perspective. Ann N YAcad Sci. 2012; 1250:73-79.
4. Madkaikar M, Mishra A, Desai M, Gupta M, Mhatre S, Ghosh K. Comprehensive report of primary immunodeficiency disorders from a tertiary care center in India. J Clin Immunol. 2013; 33(3):507-512.
5. Chinnabhandar V, Yadav SP, Kaul D, Verma IC, Sachdeva A. Primary Immunodeficiency Disorders in the Developing World: Data from A Hospital-Based Registry in India. Pediatric Hematology and Oncology.2013 June12 (Epub ahead of print) (doi:10.3109/08880018.2013.805346)