REFERENCES

1. Agarwal BR, Gurrimbhoy Z. Resolution of cyclic neutropenia by intramuscular gamma globulin in a case of common variable immunodeficiency with predominantly antibody deficiency. Indian Pediatr 1994, 31: 320-322.

2. Guerry D, Omine M, Perry S, Wolff SM. Periodic hematopoiesis in human cyclic neutropenia. J Clin Invest 1973, 52: 3220.

3. Lukens JN. Immune deficiency disease: Inherited and acquired. *In:* Wintrobes Clinical Hematology, 9th edn. Eds Lee GR, Bithell TC, *et al.* Philadelphia, Lea and Febiger, 1993, pp 1676-1702.

4. Wright JJ, Wagner DK, Blaese RM, Hagengruber C, Waldmann TA, Fleisher TA. Characterization of common variable immunodeficiency: Identification of a subset of patients with distinctive immunophenotypic and clinical features. Blood 1990, 76: 2046-2051.

Reply

The term cyclic hematopoesis is more appropriate-yes, we would agree. However, due to the relatively long half life of red cells and platelets compared with neutrophils, anemia and significant thrombocytopenia are not seen and are difficult to document(l). Since this was not documented in our case we preferred to use the term cyclic neutropenia.

This child had no organomegaly. The negative findings were not mentioned to keep the case report short. Splenectomy is without benefit for cyclic neutropenia and common variable immunodeficiency (CVID). Anyway, we would certainly hesitate to consider splenectomy for a $1^{1/2}$ year old child due to the obvious risk of infective complications in a young immunocompromized infant.

CVID syndrome describes a highly heterogenous group of patients with hypogammaglobulinemia, decreased ability to produce antibodies after antigenic stimulation and variable degrees of T-cell impairment(2). Patients with CVID can present in infancy and childhood, or during the second and third decades of life. B-cell numbers can be normal, increased or absent. T-cell analysis in some patients demonstrates decreased numbers of CD3 and CD8 cytotbxic/suppressor T-cells leading to an *increased* CD4:CD8 ratio(3). Since T-lymphocyte subsets were not studied in this case, this discussion would be redundant.

This child had isolated neutropenia with the Hb and platelet counts being normal and the antiplatelet antibodies and Coomb's test were negative. We would disagree with Parikh that a persistent viral infection could not explain cyclic neutropenia. In fact, recurrent granulocytic aplasia as clinical presentation of a persistent parvovirus B19 infection has been reported(4). Parvovirus B19, with minor genetic alterations or other similar viruses might be cytotoxic not only to erythroid progenitor cells but also to other hematopoetic progenitor cells. This may result in dysregulation of myeloid

progenitor cells with exaggerated cyclic oscillations in the rate of bone marrow production.

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REFERENCES

1. Gibson BES. Inherited disorders. *In:* Fetal and Neonatal Hematology. Eds Hann IM, Gibson BES, Letsky EA. London, Bailiere Tindall, 1991, pp 241-248.
2. Strobel S. Non-malignant disorders of lympocytes: Primary and acquired immunodeficiency disease. *In:* Pediatric Hematology. Eds Lilleyman JS, Hann IM. Edinburgh, Churchill Livingstone, 1992, pp 291-293.
3. Cunningham-Rundles C. Clinical and immunologic analysis of 103 patients with common variable immunodeficiency. J Clinlmm 1989,9: 22-23.
4. Pont J, Puchhammer-Stockl E, Chott A, *et al.* Recurrent granulocytic aplasia as clinical presentation of a persistent parvovirus B19 infection. Br J Hematol 1992, 80: 160-165.

Hypokalemic Muscle Paralysis

Dandge *et al*.(l) report the case of an 11-year-old boy with recurrent attacks of hypokalemic muscle paralysis since 3_k years of age. The patient had marked failure to thrive with weight less than the third percentile for the age. The serum potassium levels were very low ranging from 1.7-2.2 mEq/L. However, a diagnosis of primary hypokalemic periodic paralysis, an extremely rare condition, in this patient raises some concerns.

Firstly, before making this diagnosis other causes of hypokalemic paralysis must be carefully excluded. Episodic hypokalemia starting early in life is an important feature of renal tubular acidosis (RTA)(2). Recurrent epidoses of hypokalemia may also occur in hyperaldosteronism, hyperthyroidism, Bartter's syndrome and villous adenoma of the colon. In all of the latter conditions systemic acidosis is absent(3). The diagnosis of hypokalemic periodic paralysis is one of exclusion, and most patients with this condition have a family history of a similar disorder. The usual age of onset in more than 90% cases is between 7 and 21 years and onset of symptoms before 5 years of age is most unusual(4).

This patient at admission had a blood pH of 7.34 and bicarbonate level of 14.6 mEq/L. In the presence of failure to thrive and episodic hypokalemia since early childhood, these values are highly suggestive of distal RTA. An accurate measurement of the early morning urinary pH and following oral administration of ammonium chloride was essential in this patient. The presence of an inappropriately alkaline urine (pH more than 6) in patients with systemic