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Letter to Editor

Hematological Data of Cystic Fibrosis Patients in a Tertiary Care Centre in Saudi Arabia

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Dear Sir,

The incidence of Cystic fibrosis (CF) in Saudi Arabia was reported to be 1 in 4243 children[1]. Epidemiological and genetic data have been described in details in many Gulf countries[2-4], but no hematological data has been described before in the Arabian countries. In this report we present the hematological data of the largest CF population in the Gulf area and discuss its relation to mortality.

One hundred and eighty eight Cystic Fibrosis (CF) patients were diagnosed during the period November 1992-November 2002. One hundred and sixty two CF patients are alive. Ninety-eight (52%) were males and 90 (48%) were females. Age at diagnosis was 2.9 ± 3.5 years and mean follow up period was 3.2 ± 2.8 years. One hundred and eighty patients (96%) had pancreatic insufficiency. The median survival was calculated to be 10 years[3]. Initial blood tests that were drawn at diagnosis showed: Hb level was 116 ±15 (normal 132-17 g/ L), HCT 0.244 ± 123 (N 0.38-0.52 Ratio), MCV 192 ± 13 (N 80-94 fl), MCHC 333 ± 9 (N 290-370 g/L), MCH 26 ± 3 (N 27-32 pg), Platelet count 412 ± 137 (10 ^ 9/L), Retic 79 ± 30 (N 25-85 10 ^ 9/L), Fe level 11 ± 6 (N 6-24 umol/L), TIBC 64 ± 20 (N 19.7-60.2 umol/L), Transf 0.143 ± 0.09 (N 2.1-3.0 g/L), Vit E 19 ± 12 (N 14-44 umol/L), total protein level 67 ± 12 (N 65-81g/L) and Alb 40 ± 0.5 (N 35-50 g/L). There is no significant difference between males and females in all-hematological values. Comparison between hematological values of CF patients who died and those who are alive have shown that high HCT, low MCHC, low MCV and low albumin levels were lower in those who died compared to those who are alive during this report (P values= 0.0002, 0.0002 and 0.001 respectively) (Table 1). Hemoglobin electrophoresis was done in 20 patients who presented with anemia. It showed 10 patients were carrier for sickle cell disease, and one patient with homozygous sickle cell disease which was confirmed by DNA analysis. One patient had hemolytic anemia due to Vitamin E deficiency and developed gall bladder stones at 5 months of age. Glucose 6-phosphate dehydrogenase test was done on 10 patients and was positive in 2 of them.

Anemia in CF patients has been described before as presenting symptoms. Dolan and Wilfond[5,6] described few cases with hemolytic anemia, hypoalbuminemia and edema during early presentation of CF patients and assumed vitamin E deficiency causes hemolysis due to susceptibility of the erythrocyte membrane to oxidant stress and the edema is due to hypoalbuminemia. Most of our patients have shown normal

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hemoglobin with hypochromic normocytic picture. Further follow up of these patients has shown bad prognosis for those patients who develop high hematocrit, low MCV, low MCHC and low albumin in the presence of normal hemoglobin. One hypothesis to explain a false high hemoglobin level in the presence of anemia may be that it is due to increase in erythropoietin activity secondary to chronic infection and malnutrition[7,8] (Table 1).

Table 1. **Comparison of Hematological values in CF patients who died and those who are alive**

Variable	Alive	Died	P value
Hemoglobin level	115 ± 18	121 ± 17	0.2
Hematocrit	0.224 ± 0.12	0.350 ± 0.07	0.0002
Mean corpuscular volume	214 ± 138	76 ± 7	0.0002
Mean corpuscular hemoglobin Concentration	334 ± 8	328 ± 9	0.01
Mean corpuscular hemoglobin	26 ± 3	25 ± 3	0.11
Platelet count	402 ± 126	463 ± 182	0.1
Reticulocyte count	82 ± 31	68 ± 32	0.5
Iron level	12 ± 5	6 ± 4	0.12
Total iron binding capacity	64 ± 22	63 ± 9	0.9
Transferrin	0.16 ± 0.1	10 ± 0.04	0.3
Vitamin E level or alpha tocopherol	19 ± 12	19 ± 18	0.9
Total Serum Protein	65 ± 11	70 ± 11	0.1
Albumin level	40.8 ± 0.5	35 ± 1	0.001

In conclusion: routine follows up of hematological indices to detect early signs of anemia should be done frequently in CF patients. High HCT, low MCV, low MCHC and low albumin are factors related to poor prognosis and early death in CF patients. Iron supplement in conjunction with Vitamin E should be given to CF patients even in the presence of normal hemoglobin.

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