RESULTS

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This study included 100 thalassemia major children and 25 healthy ones as a control group. The thalassemic patients were divided into 2 groups according to the regularity in desferal in take : 25 cases take the drug regularly by the subcutaneous infusion pump in a dose of 25 mg/kg daily over 8 hours for 3-5 days each week. The other 75 cases were on irregular therapy because they could not get the expensive pump. They take the same dose of desferal i.e. 25 mg/kg slowly intravenously over 2 hours in 50 cc of 5% glucose solution just after transfusion of 10 cc/kg packed cells every 2-5 weeks.

A full clinical evaluation of each case was done including a thorough history and full clinical examination. Splenectomy was done for 13 cases out of the 75 irregular ones and for 19 cases out of those 25 on more regular therapy. Pneumococcal vaccine was given for all splenectomized cases as well as long acting penicillin. Hepatitis B vaccine with calcium and vit. C are given routinely. All thalassemic cases, being of the major variety, present their illness for the first time during the first year of life when they received their first blood transfusion. The frequency of transfusions depends mainly on the compliance of the patients more than the availability of blood. "The Thalassemia Federation" in Egypt which is a branch of the international one tries hardly to make blood, desferal, vaccines, long acting penicillin and other currently used drugs available freely for these patients to lower the coast of this life long therapy which cannot be afforded by many patients.

Hemoglobin electrophoresis was done for each case. It is noted that the mean values of Hb% in thalassemic children is far below the goal of ideal therapy (5.65 ± 1.0 gm% in the irregular group and 6.16 ± 1.2 gm% in the regular one).

Estimation of the serum level of Fe, Al, Pb, Ca, Mg, Zn, Cu and Mn is done by the spectrophotometer. Five cc of blood is withdrawn from the needle in which blood will be given just before blood transfusion. Clinical data and lab results for cases and controls were put in tables were cases number 1 to 75 represent the irregular therapy group, cases from 76 to 100 are those on more regular therapy and from 101 to 125 are the healthy control group.

The mean values of serum iron, being not a sensitive indicator particularly in the regularly treated thalassemic children, was below expected values with significant effect of regular desferal administration. Aluminum levels on the other hand, although significantly affected by desferal therapy, they did not show a significant effect of regularity in administration. The effect of the disease and possibly the drug on serum level of Ca, Mg, Zn and Cu has been veiled in our study by the routine Ca administration and frequent transfusions. The effect on Pb an Mn. has to be re-evaluated ANOVA test between the mean values of each elements in the 3 groups together and the significance of differences between each 2 groups is shown in Tables 1-8 and Figs. 1-8. The significant correlations between variants in each group particularly the irregular one is shown in Figs. 9-14. The significance of splenectomy effect on variants in the 2 thalassemic groups is also shown in tables 9-10.
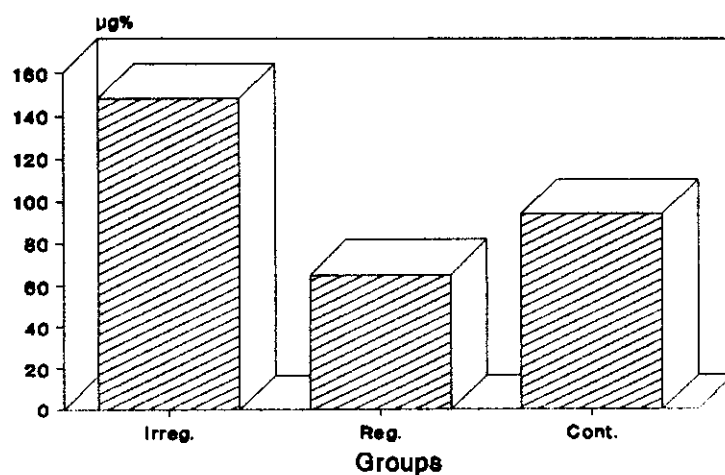
Table (1): Differences in serum Iron (Fe).

Group	Irreg.	Reg.	Control
Number (n)	75	25	25
Mean (μg%)	147.93	65.00	93.60
$\pm$ S.D.	$\pm$ 60.66	$\pm$ 8.78	$\pm$39.70

ANOVA test between the 3 groups shows v.highly sig. differences at $P < 0.0001$.

Differences between each 2 groups show

- Irreg x Reg. P = 0.0001   i.e. v. highly significant.

- Irreg. x Cont. P = 0.001   i.e. highly significant.

- Reg. x Cont. P = 0.01   i.e. significant

Fig.(1): Mean serum (Fe) levels.

Table (2): Differences in serum Aluminum (Al).

Group	Irreg.	Reg.	Control
Number (n)	75	25	25
Mean (µg%)	0.52	0.56	0.71
± S.D.	± 0.10	± 0.08	± 0.11

ANOVA test between the 3 groups shows v.highly sig. differences at $P < 0.0001$.

Differences between each 2 groups show

- Irreg x Reg. P = 0.01   i.e. not significant.

- Irreg. x Cont. P = 0.0001   i.e. v.highly significant.

- Reg. x Cont. P = 0.001   i.e. highly significant
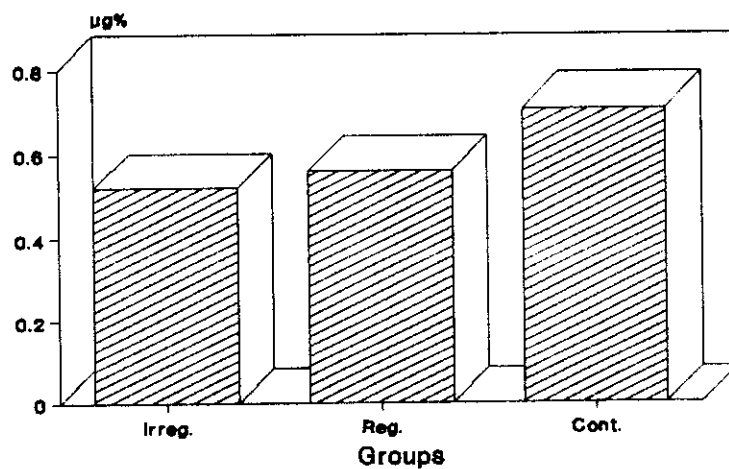
Fig.(2): Mean serum (Al) levels.
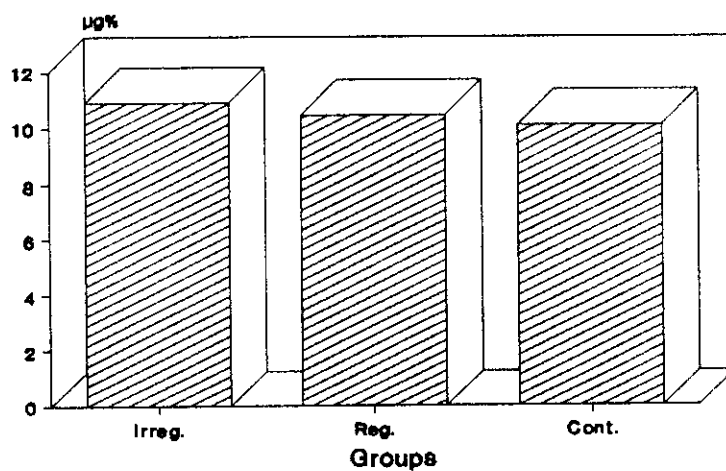
Table (3): Differences in serum Lead (Pb).

Group	Irreg.	Reg.	Control
Number (n)	75	25	25
Mean (µg%)	10.92	10.45	10.10
± S.D.	± 5.62	5.80	6.14

ANOVA test between the 3 groups shows insignificant differences.
Differences between each 2 groups show

- Irreg x Reg. P > 0.1   i.e.not significant.

- Irreg. x Cont. P > 0.1   i.e. not significant.

- Reg. x Cont. P > 0.1   i.e. not significant
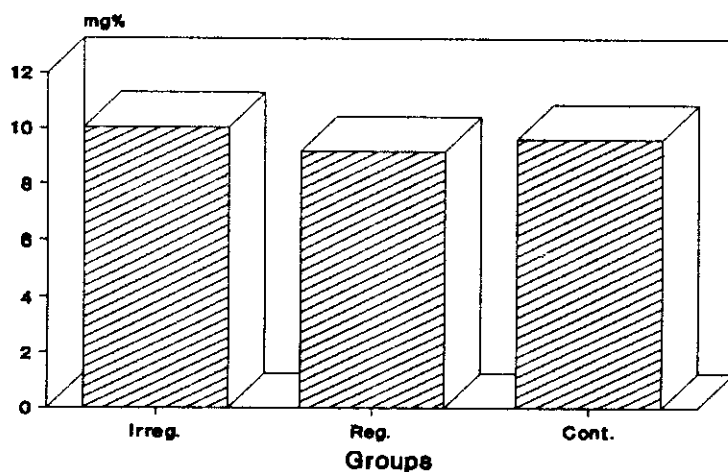

Fig.(3): Mean serum (Pb) levels.

Table (4): Differences in serum Calcium (Ca).

Group	Irreg.	Reg.	Control
Number (n)	75	25	25
Mean (mg%)	10.06	9.19	9.64
± S.D.	± 1.52	± 1.56	± 0.93

ANOVA test between the 3 groups shows significant differences at $P = 0.01$.

Differences between each 2 groups show

- Irreg x Reg. $P = 0.02$ i.e. significant.

- Irreg. x Cont. $P = 0.2$ i.e. insignificant.

- Reg. x Cont. $P = 0.1$ i.e. insignificant

**Fig.(4): Mean serum (Ca) levels.**

Table (5): Differences in serum Magnesium (Mg).

Group	Irreg.	Reg.	Control
Number (n)	75	25	25
Mean (mg%)	1.71	2.20	1.57
± S.D.	± 0.60	± 0.67	± 0.53

ANOVA test between the 3 groups shows v.highly sig. differences at $P < 0.0005$.

Differences between each 2 groups show

- Irreg x Reg. $P = 0.01$ i.e.   highly significant.

- Irreg. x Cont. $P = 0.3$ i.e. not significant.

- Reg. x Cont. $P = 0.001$ i.e. v. highly significant.
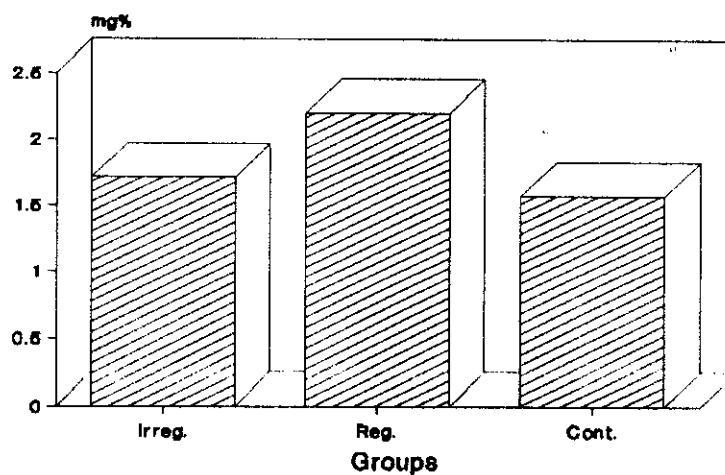
Fig.(5): Mean serum (Mg) levels.
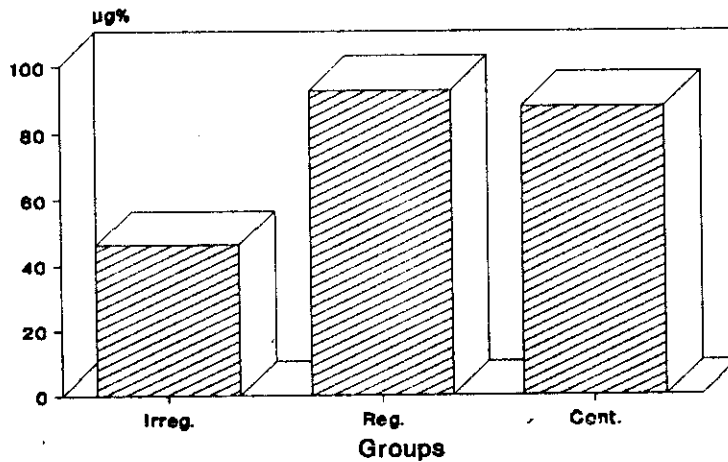
Table (6): Differences in serum Zink (Zn).

Group	Irreg.	Reg.	Control
Number (n)	75	25	25
Mean (µg%)	46.11	92.76	87.88
± S.D.	± 19.45	± 22.54	24.11

ANOVa test between the 3 groups shows v.highly sig. differences at $P < 0.0001$.

Differences between each 2 groups show

- Irreg x Reg. P = 0.0001   i.e. v. highly significant.

- Irreg. x Cont. P = 0.0001   i.e. highly significant.

- Reg. x Cont. P = 0.5   i.e. not significant

Fig.(6): Mean serum (Zn) levels.
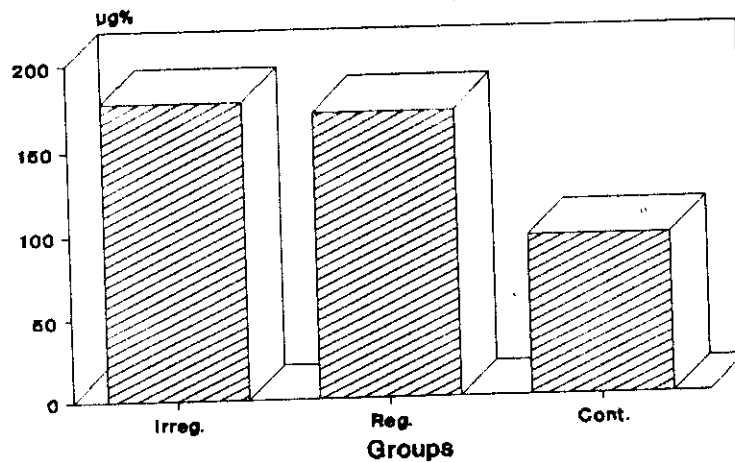
Table (7): Differences in serum Copper (Cu).

Group	Irreg.	Reg.	Control
Number (n)	75	25	25
Mean (μg%)	177.51	171.24	96.6
± S.D.	± 12.93	± 13.73	± 10.30

ANOVA test between the 3 groups shows v.highly sig. differences at $P < 0.0001$.

Differences between each 2 groups show

- Irreg x Reg. P = 0.05   i.e.   not significant.

- Irreg. x Cont. P = 0.0001   i.e. v.highly significant.

- Reg. x Cont. P = 0.0001   i.e. v.highly significant
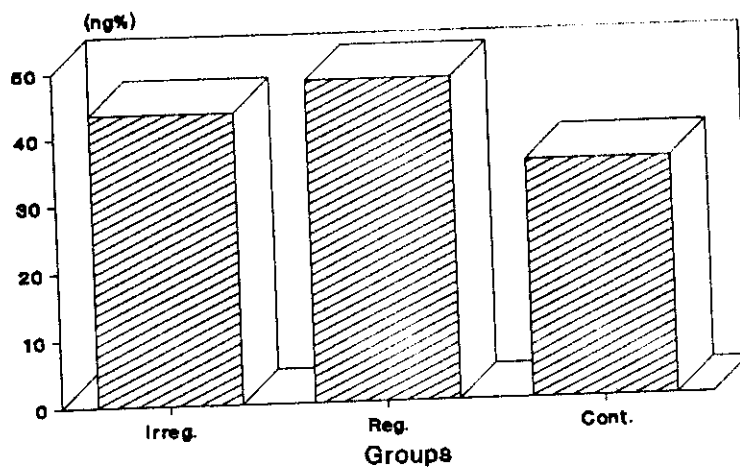

Fig.(7): Mean serum (Cu) levels.

Table (8): Differences in serum Manganese (Mn).

Group	Irreg.	Reg.	Control
Number (n)	75	25	25
Mean (ng%)	43.65	48.32	35.52
± S.D.	± 18.18	± 16.99	± 9.90

ANOVA test between the 3 groups shows v.highly sig. differences at $P = 0.02$.

Differences between each 2 groups show

- Irreg x Reg. $P = 0.03$ i.e.not significant.

- Irreg. x Cont. $P = 0.05$ i.e. significant.

- Reg. x Cont. $P = 0.01$ i.e. significant

Fig.(8): Mean serum (Mn) levels.

Effect of splenectomy

Table (9): Irregular group [Number of splenctomized cases = 13, non -splenctomized cases = 62, (df = 73)].

	Fe	Al	Pb	Ca	Mg	Mn	Zn	Cu	HbA	HbA₂	HbF	Hb%
t	1.06	0.31	0.27	2.60	0.23	0.07	0.17	1.31	0.76	0.63	0.62	0.61
P	0.292	0.756	0.785	0.011	0.818	0.949	0.245	0.195	0.447	0.528	0.540	0.011

Table (10): Regular group [Number of splenectomized cases =19 , non splenectomized cases = 6 (df = 23)]

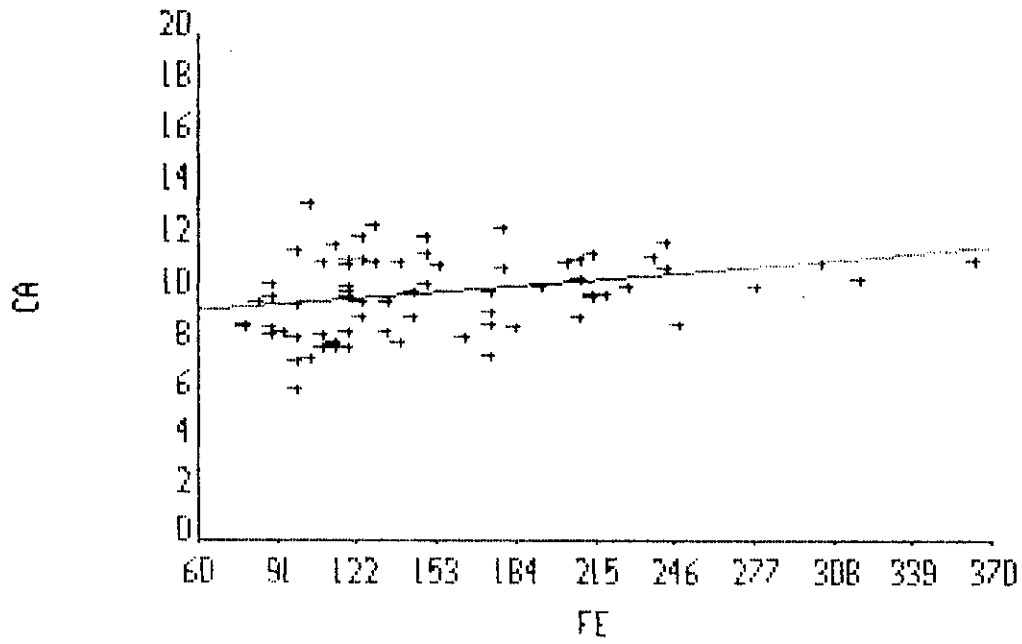|	Fe	Al	Pb	Ca	Mg	Mn	Zn	Cu	HbA	HbA₂	HbF	Hb%
t	0.49	1.86	0.26	0.15	1.60	0.6	1.94	0.73	2.11	1.91	1.19	1.17
P	0.631	0.076	0.796	0.882	0.124	0.557	0.064	0.476	0.046	0.069	0.247	0.254

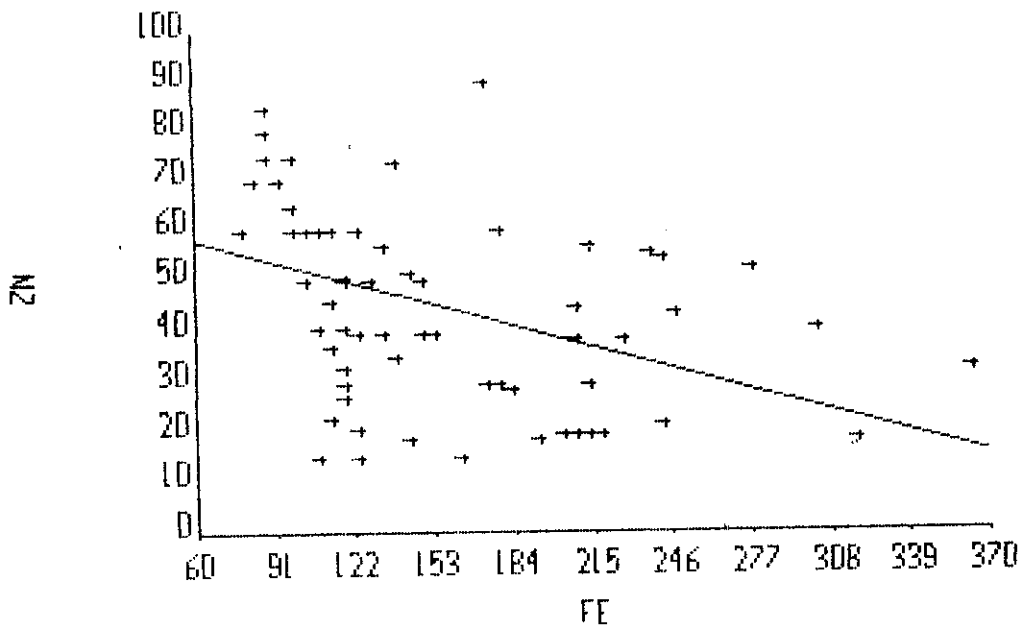**Fig. (9):** Correlation between Fe and Ca in the irregular group.

Fig. (10): Correlation between Fe and Zn in the irregular group.

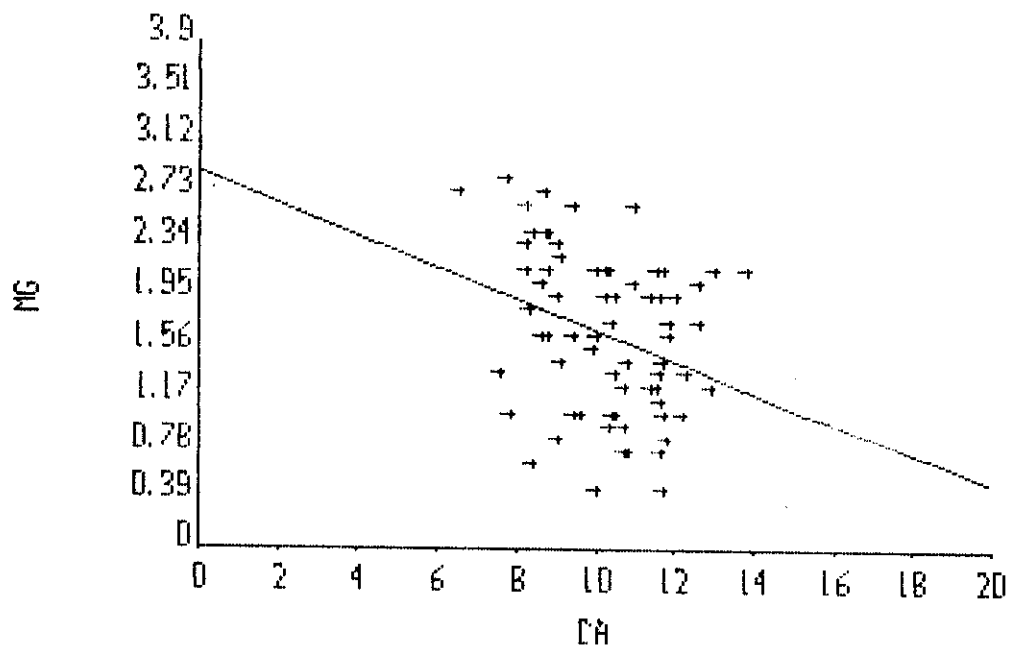**Fig. (11):** Correlation between Ca and Mg in the irregular group.
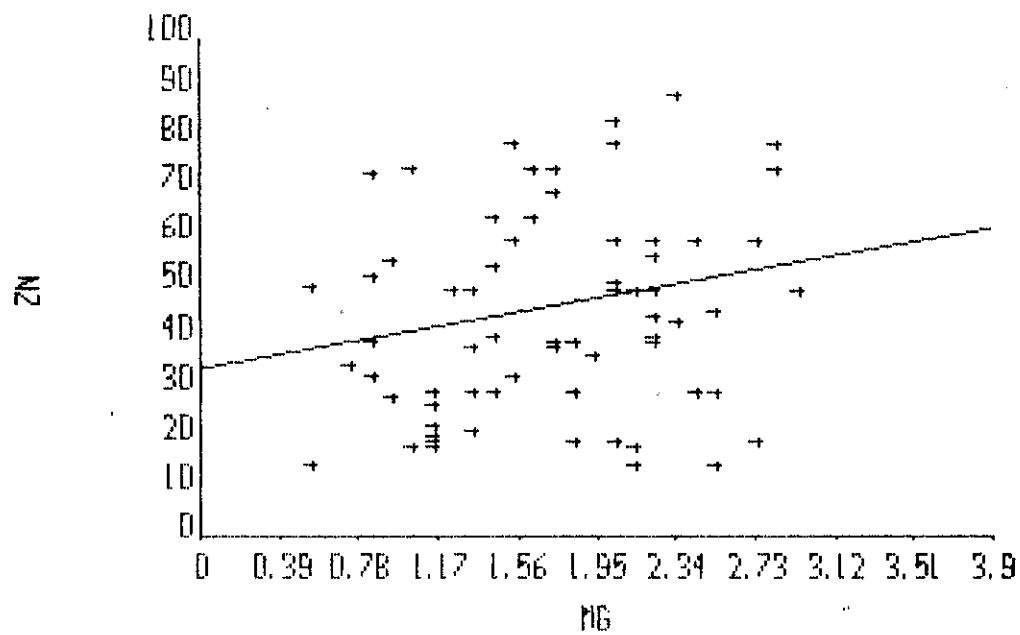

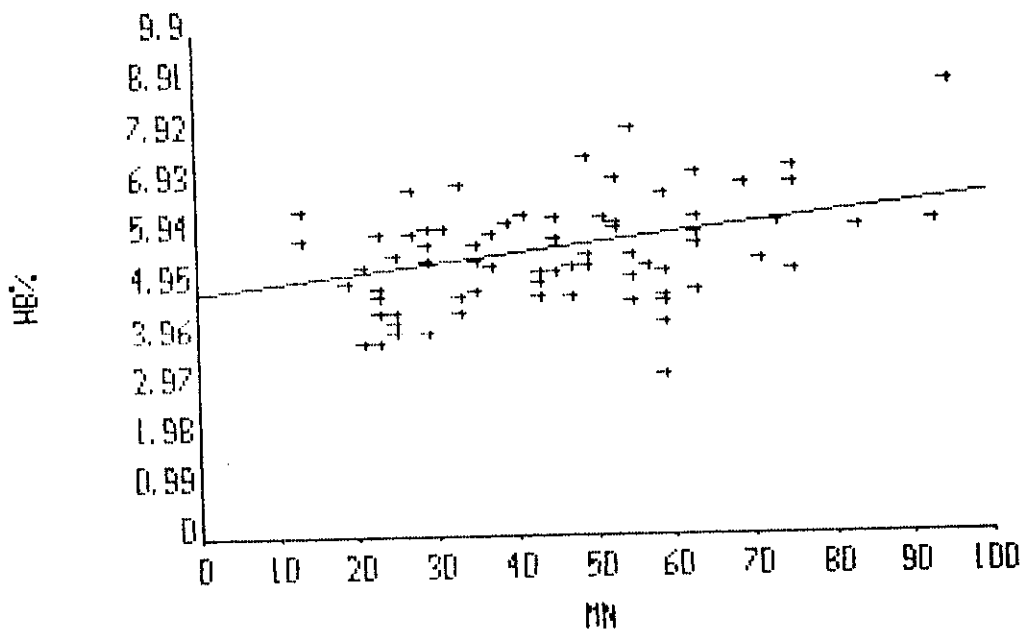**Fig. (12):** Correlation between Mg and Zn in the irregular group.

Fig. (13): Correlation between Mn and Hb% in the irregular group.