

Usefulness of monitoring Vit D in patients with inborn errors of metabolism on protein restricted diet

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Introduction : Vitamin D is very important. It is responsible for bone development, Calcium absorption from intestine etc. Its deficiency increases the risk of fractures and Osteoporosis. Its deficiency causes Rickets in children and osteomalacia in adults. Vit D also has important effects on the immune system, prevents cardiovascular diseases and certain cancers.¹

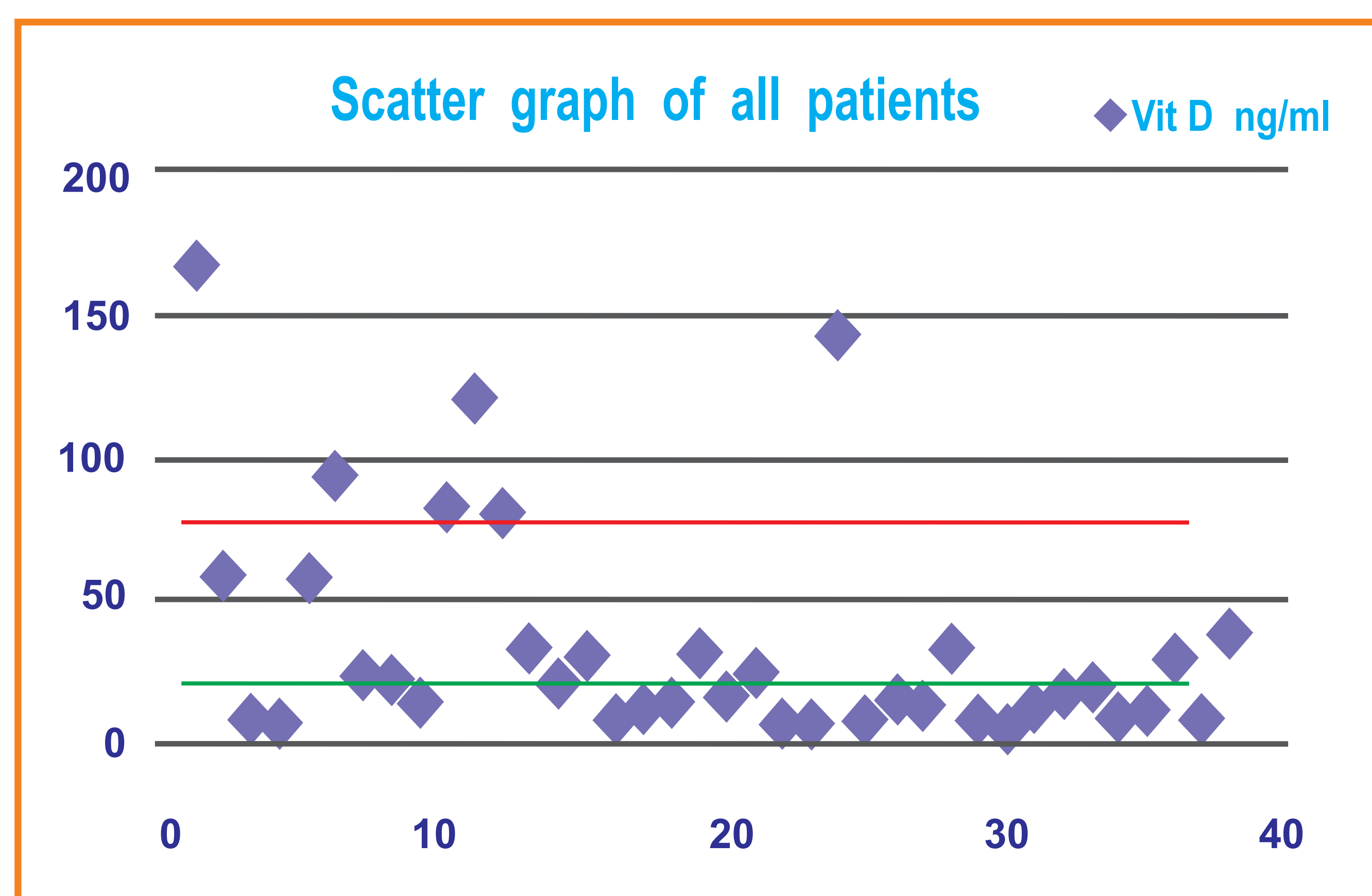
Patients suffering from Inborn Errors of Metabolism like Organic acidemia, Urea cycle defects and aminoacidopathies are on protein restricted diet. Some require amino acid supplementation and special diets. These diets contain Vit D apart from other nutrients.

Objective : To find the Vit D status in patients with inborn errors of metabolism like aminoacidopathies, Urea cycle defects and organic acidemias who are on protein restricted diet with or without dietary supplementation.

Subject : The study involves 36 diagnosed cases of IEM's on protein restricted diet who were prospectively studied from Jan 2013 to March 2014. They include amino - acidopathies ; Tyrosinemia type I (8), Phenylketonuria (4), Homocystinuria (2), Maple Syrup urine Disease (2), Urea cycle defects; Citrullinemia Type I (6), Argininosuccinic aciduria (2) and Organic acidemias; Methylmalonic aciduria (10), Propionic aciduria (1), Isovaleric aciduria (1).

Method : Serum samples were analyzed for 25 (OH) Vit (D2+D3) using IVD approved ELISA kits. Serum Calcium, Phosphorus, Alkaline phosphatase and Proteins were measured using dry chemistry. Vit D levels were analyzed in 3 subjects before and after special diet. There are not many papers providing consistent reference ranges, especially from India. Therefore we have adopted Mayo Clinic Reference range for 25 (OH) Vit D.

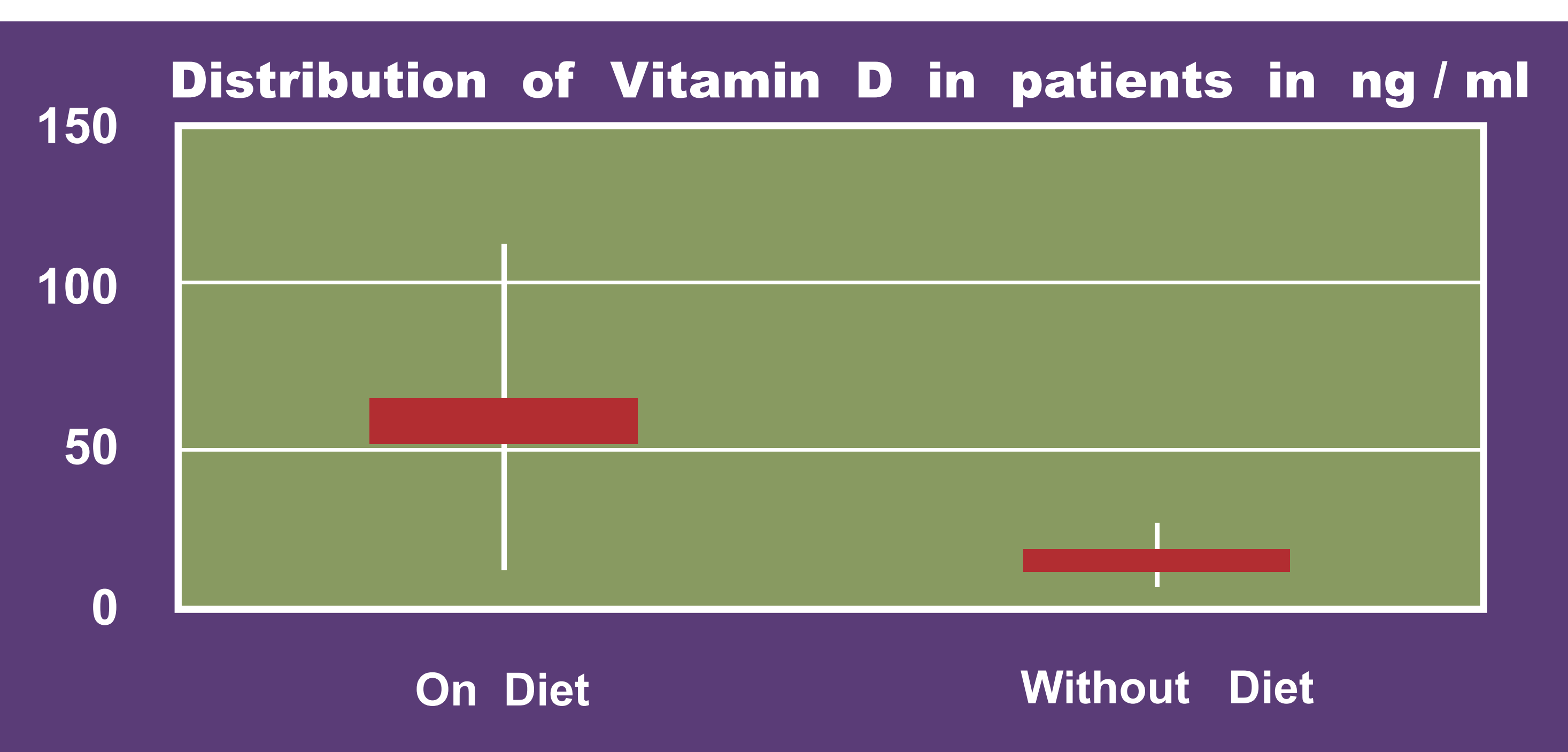
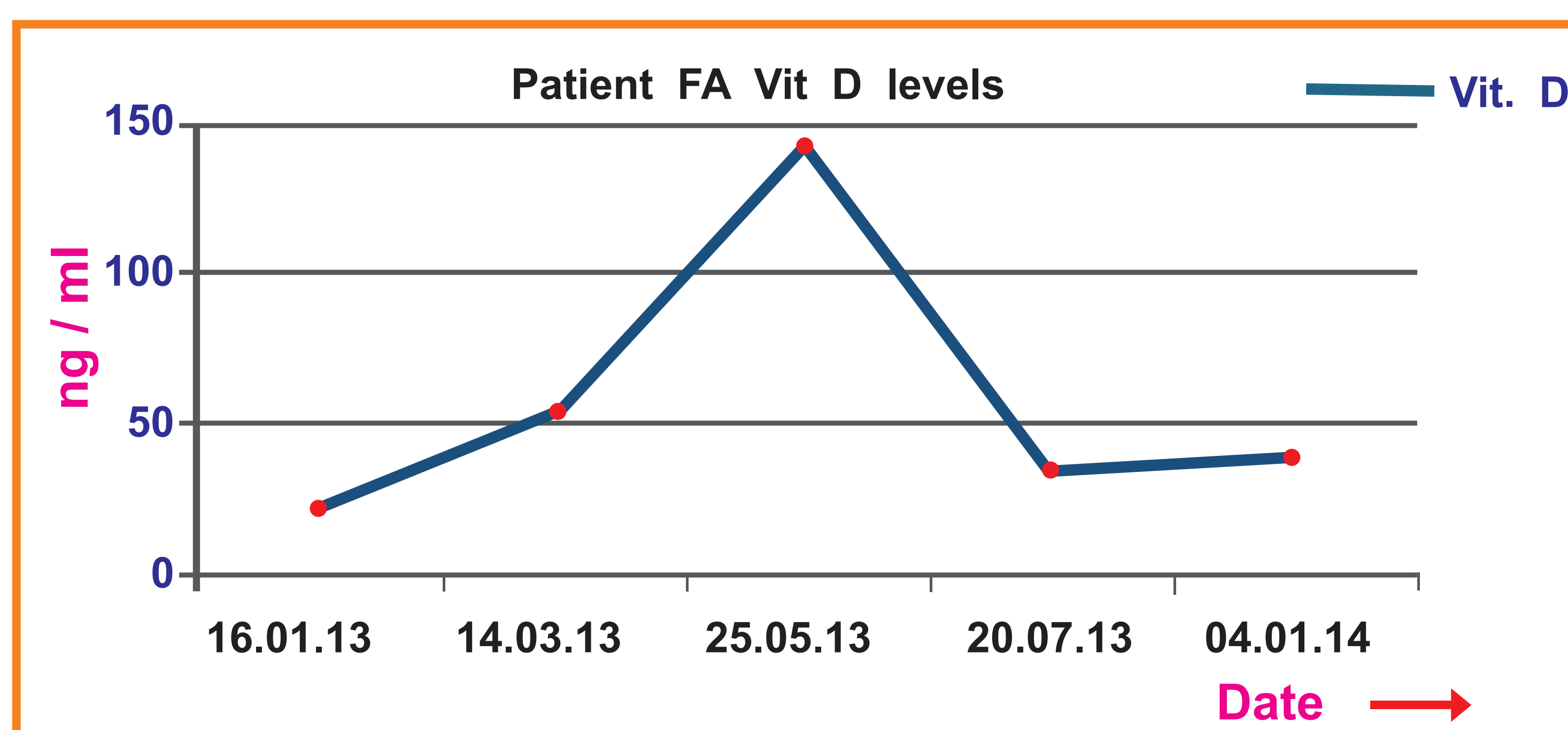
Vitamin D reference range (Mayo clinic) ²	
Deficiency	<20 ng/ml
Insufficiency	20 - 30 ng/ml
Sufficiency	>30 ng/ml
Toxicity	>80 ng/ml



Result : We found that those on special diets had a higher level of Vit D concentration (n = 15, 62.862 ± 50.456 ng/ml) with 6 being in the toxic range and 4 deficient. Those not on any special dietary supplement (n = 24, 16.320 ± 9.26ng/ml) had lower levels and 18 were deficient. Of the three patients whose readings were taken both before and after starting special diet, we found that the Vit D levels increased significantly after starting special diet. Patient FA was also on Vit D supplementation which had to be stopped after his Vit D levels reached above 140 ng/ml.

	N = 39	Mean	SD	Toxi city	Defi - cient
Patients on diet	15	62.86	50.45	6	4
Patients Not on diet	24	16.32	9.25	0	18

Patients	Before Diet	After Diet	Vit D in diet supplementation
FA	22.93	22.93	340 + 400 IU
MA	15.61	15.61	307 + 400 IU
RS	628	628	147 IU No Supp



Discussion : As far as we know there is no data pertaining to the Vit D status in children affected with inborn errors of metabolism. Harinarayan et.al has found that 50 – 90 % of population in the Indian subcontinent is Vit D deficient³. In our cohort (N=39) 75 % of Patients not taking any special diet (18/24), and 26.6 % of those taking special diet (4/15) developed Vit D deficiency. Though 40 % (6/15) of the patients receiving special diets showed Vit D levels more than 80 ng/ml, theoretically in toxic range, did not have evidence of toxicity (for eg. Hypercalcemia)

Conclusion : It is necessary to monitor Vit D concentration in patients with protein restricted diet with or without special AA supplements, as some may be deficient or may have toxic levels, especially those who are on special diets.

References :

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3. Vitamin D status in India – Its Implications and remedial measures. Harinarayan CV, Joshi SR., *J Assoc Physicicans.* 2009; 57: 40-8