

319 Vitamin K monitoring in adult CF patients

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Introduction: As well as its role in coagulation, the importance of vitamin K in bone mineralisation is becoming increasingly recognised. Although there are currently no guidelines for vitamin K supplementation or monitoring in CF patients, recent evidence indicates deficiency is common in this patient group, which also has a high incidence of bone disorders. To investigate this further, we measured vitamin K levels in a subgroup of our adult CF patients.

Method: At their routine annual screen, from a serum sample we measured vitamin K₁ and also prothrombin produced in vitamin K absence (PIVKA-II, a more sensitive measure of stored Vitamin K levels) in 16 adult CF patients (mean age 28 years [range 18 to 44]; mean FEV1 64% predicted [range 34 to 105]; 10 male).

Results: 10 patients (63%) had a low vitamin K₁ (<0.15 µg/l). Of these patients, only 1 (10%) had a BMI < 20. 2 (20%) were enzyme sufficient and only 3 (30%) had documented liver dysfunction, with prescription of ursodeoxycholic acid. PIVKA-II was not detected (i.e. < 0.20 AU/ml) in all 16 patients, indicating sufficient body stores for the production of coagulation factors.

Conclusion: Suboptimal vitamin K status was very common in our patients, as indicated by low vitamin K₁ levels. Although the absence of PIVKA-II suggests sufficient vitamin K stores for coagulation, vitamin K requirements for optimal carboxylation of osteocalcin in bone are known to be higher and stores may not be adequate to support this function. Further work needs to be carried out to assess the importance of vitamin K supplementation in adult CF patients.

320 An audit of low profile gastrostomy devices – is rupture of the internal retention balloon more common in CF?

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Following a change in manufacturer's guidelines for the care of a low profile gastrostomy device we observed 2 ruptures of the retention balloon in a CF patient: the device is then at risk of falling out and the tract closing off. We audited the use of MicKey gastrostomy buttons in our patients.

We identified 5 CF patients with a MicKey (4M/1F; median age 15.5 y (9–17.9)) and 10 non-CF patients (4M/6F; 10.7 y (3.0–16.7)) (p < 0.05) with various diagnoses, particularly neuromuscular disorders. The medical and nursing records were reviewed.

All CF patients were on long term antibiotics; 3/10 of non-CF patients. Acid suppressants were used in 3/5 CF patients and 5/10 non-CF. All CF patients were receiving pancreatic enzymes. The gastric aspirate of 14/15 patients was colonised with candida.

In the 5 CF patients we recorded 10 episodes of balloon rupture with 1 balloon intact at MicKey change. This compares to 2 balloon ruptures in the non-CF patients with 16 balloons intact (p < 0.0001 Fisher's exact test).

In the CF patients the median balloon life to bursting was 102 days (69–304); 1 CF patient's Mickey balloon was intact when changed at 110 days. In the non-CF group the 2 burst balloons had been in situ for 44 and 144 days (median 93.5) compared to a median of 82.5 days (45–280) in those not ruptured. For all patients, MicKeys with intact balloons needed changing at a median of 83 days compared to 102 days in those which had burst balloons (CI: –45 to +44; P = 0.69. Mann Whitney).

We have identified an increased rate of rupture of the retention balloon in one type of gastrostomy device in our CF patients. We now recommend to our CF patients that MicKey gastrostomy buttons are changed every 3 months or that the family receives in-depth training on the management of a displaced button.