11. Nutrition

247 Sodium depletion in infants diagnosed with CF
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Background: Oral sodium supplementation of CF infants is not standard practice in the UK [1]. Sub clinical salt depletion can result in growth impairment in CF; particularly in infancy [2]. Infant formulas and breast milk may not meet the sodium requirements of CF infants.

Method: The urinary electrolyte levels of all infants diagnosed during 2008 with CF at RHSC was measured. Spot urine analysis was measured once the infant was established on feeds. A low result is classed as a urinary sodium level less than 10 mmol/L or a sodium:potassium ratio less than 2:1. Sodium supplementation consisted of a 30% sodium chloride solution (5 mmol/sodium/l ml).

Results: In total 10 infants were diagnosed with CF, 7 on newborn screening, 2 with meconium ileus and 1 with hyperbilirubinaemia. 8 of the infants were homozygous for the DF508 mutation. 9 of the infants were pancreatic insufficient and started pancreatic enzymes. 5 of the infants were breast fed and 5 were formula fed (one with a hydrolysed formula and one a MCT formula). All infants had urinary electrolytes measured and all were found to be sodium depleted. Sodium supplementation was commenced on all infants apart from the infant who was pancreatic sufficient whose growth was on the 91st centile. The dose of sodium supplementation required to bring the urinary sodium into the normal range was between 0.6 mmol/Kg to 6 mmol/Kg. Improvement in growth was noted in 6 of the infants after commencement of supplements.

Conclusion: These results suggest infants diagnosed with CF in the UK are at risk of sodium depletion. Routine sodium supplementation should be considered in those with low urinary electrolyte levels.

Reference(s)

248 Soluble transferrin receptor for evaluating iron status in cystic fibrosis
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Background: Malnutrition in CF is commonly associated with deficiencies of iron, trace elements and fat soluble vitamins. Since CF is also associated with chronic inflammation in many patients and increasing with age, diagnosing iron deficiency (ID) might be difficult. Soluble transferrin receptor (sTfR) is a marker for iron status that is independent of inflammation.

Aim: To evaluate if more patients are diagnosed with ID when including sTfR in the annual review.

Method: During the annual review iron status and inflammatory variables are measured including hemoglobin, MCV, ferritin, transferrin saturation, ESR, CRP, haptoglobin. If 2 out of 3 results from MCV, ferritin and transferrin saturation are lower than ref. values the patient is regarded as ID ("old" definition). During 2007 sTfR was also included in the annual review. If sTfR was above ref.values and at least one of the regular measurements mentioned above was low, these patients were also regarded as an ID ("new").

Results: 134 patients (56F) with a mean age 21.6±12.7 years were included. FEVER 1.0 were 77±24% in adults and 90±17% in children. Twenty-five patients had ESR above 13 mm and 26 had CRP above 5 mg/L. sTfR was not correlated to any inflammatory variable.

Only 2/134 patients had anemia, both being ID. With the "old" definition 11 patients were ID (3F, median age 17.8, range 4.4–33.6 yrs). By adding sTfR another 2 patients were diagnosed as ID. Two out of the 6 patients with increased sTfR had no other sign of ID.

Conclusion: 11 patients (8%) of the population had ID according to the old diagnostic criteria. The addition of sTfR only recognised another 2 patients (10%). The proportion of patients with severe inflammation was relatively low which may have influenced the outcome. The use of only sTfR as indicator of ID cannot be recommended in CF.

249 Vitamin K status and supplementation in children with cystic fibrosis (CF)
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Background: Patients with CF are at risk of an impaired Vitamin K status which may lead to frank deficiency and adversely affect blood coagulation and bone formation/density.

Objectives: (1) To establish the prevalence of frank Vitamin K deficiency and suboptimal Vitamin K status in children with CF. (2) To evaluate the effectiveness of age related doses of Vitamin K on a weekly basis to correct a suboptimal Vitamin K status and a daily basis for frank deficiency.

Patients and Methods: 31 children with PI CF and 2 children with pancreatic sufficient (PS) CF had base line data collected for serum PIVKA II and serum Vitamin K1. Children with a suboptimal Vitamin K1 (Vitamin K1 <0.15ug/l) received an age related weekly dose of Phytomenadione (Vitamin K1) (10mg for those over 2 years (n = 32) and a 5 mg for those under 2 years (n = 1)). Children with frank Vitamin K deficiency (raised PIVKA II >0.2au/ml) received a daily dose of 10 mg Phytomenadione (n = 5, all over 2 years of age).

After 1 year the baseline blood tests were repeated. None of the children had previously received Vitamin K supplementation.

Results: Sixteen children (48%) had a compromised Vitamin K status. Of these patients 5 (15%) had frank Vitamin K deficiency and 11 (33%) had a suboptimal Vitamin K1 level.

Administration of Phytomenadione (Vitamin K1) as per the protocol corrected all the children with either frank Vitamin K deficiency or suboptimal Vitamin K status.

Conclusion: Children with PI CF are more likely to have a suboptimal Vitamin K status than a frank deficiency and this can be corrected with an age related weekly dose of Vitamin K.

348 Vitamin K deficiency in CF patients is frequent despite its regular supplementation
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Cystic fibrosis (CF) patients are at high risk for developing vitamin K deficiency and are commonly supplemented with this vitamin. Since methods dedicated for monitoring vitamin K status are hardly available, the data related to the efficacy of the supplementation are sparse. Therefore, we have aimed to assess vitamin K status of our routinely supplemented patients.

Material and Methods: The study comprised 42 CF patients aged from 3 to 33 years supplemented at least once a week. Vitamin K status was assessed by the measurement of PIVKA II concentrations (ELISA, Roche Diagnostic, Poland).

Results: In 30 patients PIVKA II concentrations were lower than detection limit provided by the manufacturer (<4ng/ml), suggesting normal vitamin K status. In 12 (28.6%) subjects this undercarboxylated protein was detectable (range of 5 to 97 ng/ml).

Conclusions: Despite vitamin K supplementation its deficiency in CF patients is still frequent.