

Department of Paediatric Respiratory Medicine and Cystic Fibrosis

# Vitamin supplementation in Cystic fibrosis

## **Vitamin D supplementation**

The CF trust has recently produced new guidelines for vitamin D supplementation and bone health in children with CF (see CF Trust website for more details).

#### Metabolism

Vitamin D3 (colecalciferol) is either absorbed in the diet or produced in the skin by photoconversion of 7-dehydrocholesterol. D3 It is converted by the liver to 25(OH)D3, which is the major form of vitamin D circulating in the blood. 25(OH)D3 is converted into 1,25(OH)D3 and 24,25(OH)D3 in the kidney. These two forms are active and serve to increase intestinal absorption of calcium.

Vitamin D2 (ergocalciferol) is not naturally produced by vertebrates, but is obtained in the diet, and is often given as a Vitamin D supplement.

Recommended first line vitamin D supplementation is with Dalivit, Abidec or Vitamin A+D capsules. These are Vitamin D2 supplements. Vitamin D3 supplements can also be given. Preparations available on the UHW formulary include Adcal D3 dissolve, Calcichew D3 caplets, Fultium 800 IU capsules (recently licensed in UK), ZymaD (10,000 IU / ml (licensed in France) and Dekristol 20,000 IU capsules (licensed in Germany).

## Recommended supplementation

Recommended daily doses of ergocalciferol (D2) or colecalciferol (D3) for routine use in pancreatic insufficient children:

Age	Dose / day
Birth – 1 year	400 IU
1-12 years	400-800 IU
>12 years	800-2000IU

1 microgram of vitamin D is equivalent to 40 IU

#### Suitable Vitamin preparations for uncomplicated therapy

Preparation	Vitamin D (IU)	Vitamin A (IU)	
Abidec (0.6 mls)	400	1333	<b>CONTAINS PEANUT OIL</b>
Dalivit (0.6 mls)	400	5000	
A+D capsule	400	4000	
Multivitamins BPC	300	2500	

#### Vitamin D alone & combined Vitamin D / Calcium supplements for more complicated cases

Preparation	Vitamin D (IU)	Calcium
Adcal D3 Dissolve	400	15 mmol
Calcichew D3 caplets	400	12.5 mmol
Fultium D3	800	CONTAINS PEANUT OIL
ZymaD (10 ml bottle)	10,000 / ml	
Dekristol D3 capsules	20,000 / capsule	CONTAINS PEANUT OIL

## Management of low vitamin D levels

## Recommended total levels of 25(OH)D are **30-60µg/I** (75-150nmol/L).

We measure Vitamin D levels and PTH levels at annual review, together with calcium, phosphate and alkaline phosphatase. Vitamin D results are usually given for both 25(OH)D2 and 25(OH)D3. There is strong circadian variation to PTH levels and a relationship to eating. Early morning levels taken after a small breakfast will be higher than afternoon levels taken after a large lunch.

## FOR CHILDREN WHO CAN SWALLOW TABLETS (use Dekristol capsules)

### If total serum 25(OH)D is above 30 µg/l and PTH is normal

• No action

### If PTH is high:

- Serum 25(OH)D level 20-30 μg/l; give 40,000iu colecalciferol D3 once weekly for 4 weeks
- Serum 25(OH)D level <20  $\mu$ g/l; give 40,000iu colecalciferol D3 once weekly for 6 weeks
- Repeat 250HD and PTH after 3 months

#### If PTH is normal:

- Serum 25(OH)D level 20-30 μg/l; give one dose 40,000iu colecalciferol D3
- Serum 25(OH)D level <20 μg/l; give 40,000iu colecalciferol D3 once weekly for 2 weeks
- Serum 25(OH)D level <10 nmol/L; give 40,000iu colecalciferol D3 once weekly for 4 weeks
- Repeat 25(OH)D and PTH after 6 months

#### FOR CHILDREN WHO CANNOT SWALLOW TABLETS (use ZymaD liquid)

## If total serum 25(OH)D is above 30 μg/l and PTH is normal

No action

#### If PTH is high:

- Serum 25(OH)D level 20-30 μg/l; give 20,000iu colecalciferol D3 once weekly for 4 weeks
- Serum 25(OH)D level <20  $\mu$ g/l; give 20,000iu colecalciferol D3 once weekly for 6 weeks
- Repeat 25OHD and PTH after 3 months

### If PTH is normal:

- Serum 25(OH)D level 20-30 μg/l; give one dose 20,000iu colecalciferol D3
- Serum 25(OH)D level <20 μg/l; give 20,000iu colecalciferol D3 once weekly for 2 weeks
- Serum 25(OH)D level <10 nmol/L; give 20,000iu colecalciferol D3 once weekly for 4 weeks</li>
- Repeat 25(OH)D and PTH after 6 months

#### **THEREAFTER:**

- Check compliance. Remind the family that all fat soluble vitamins should be given with fat containing food and Creon.
- Increase dose of vitamin D up to a maximum of 2000iu per day as required
- Dietician should make an assessment of calcium intake
- Take care with combined preparations such as Abidec, Dalavit and Vit A+ D capsules such that
  vitamin A levels don't exceed the upper limit of normal. Maximum recommended daily doses
  of vitamin A are not established but 10,000 IU has been suggested. High Vitamin A doses are
  teratogenic and teenage girls of reproductive age should definitely not be on a vitamin A dose
  > 10,000 IU / day.

• We recommend the maximum dose of combined A+D treatment should not exceed

Dalivit 1.8 mls
 Abidec 3mls
 Vitamin D 1200IU; Vitamin A 15,000 IU
 Vitamin D 2000IU; Vitamin A 6500 IU
 Vitamin A+D 3 capsules/ day
 Vitamin D 1200IU; Vitamin A 12,000 IU

- If vitamin D levels are still low on the above maximum dosing, introduce a vitamin D only supplement such as Fultium D3. If calcium intake is low use a combined Vitamin D / calcium preparation such as AdcalD3 dissolve or calcichew D3 caplets. These dissolvable preparations will be most suitable for younger children.
- With greater problems of malabsorption, calcitriol is very well absorbed since it is hydroxylated - 25(OH)D levels will not reflect calcitriol therapy and so other approaches including regular serum and urinary calcium measurements are needed for surveillance and monitoring.

## **Vitamin E supplementation**

Age	Dose /day
Birth-12 months	50 mg
1-10 years	100mg
>10 years	200mg

#### **Formulations**

- Vitamin E suspension 100mg/ml.
- Vita E capsules come as 75IU, 200IU and 400IU
- Solubel Vitamin E preparation do exist and may be used rarely in difficult cases

Children older than 5 years who are able to swallow tablets can generally be put on one 200IU capsule per day.

1mg of vitamin E is equivalent to 1-1.5 IU depending on the exact preparation

# **Vitamin K supplementation**

Vitamin K is only routinely required in children with CF related liver disease where PT is prolonged. Standard recommended dose: 10mg/day for children and adults.

Formulation: Menadiol phosphate 1 tablet = 10mg.

Children with CF undergoing major surgery should receive a dose of vitamin K IV before surgery irrespective of their clotting status, as consumption of clotting factors as a consequence of surgery may unmask a relative deficiency.