What are the extra vitamin requirements for Cystic Fibrosis (CF) patients?

The Royal Brompton Hospital and Oxford University Hospital are our local tertiary CF centres serving Buckinghamshire; but many patients also visit paediatricians at Bucks Healthcare Trust, where this is more convenient, for follow up appointments.

In CF the fat soluble vitamins may be lost in stool, due to the difficulty absorbing fats. The aim is to keep vitamin levels in the upper limit of the normal range. The idea behind vitamin replacement is to ensure that individuals do not become depleted, in order to keep their levels within the normal range.

Patients with CF will have annual blood tests to check Vitamin A, D & E status, and clotting/LFTs to assess the need to replace vitamin K. The secondary care specialist will make an individualised recommendation based on these results.

*Usual Recommended Daily Amounts in CF are shown in the table:*

<table>
<thead>
<tr>
<th>Age</th>
<th>Vitamin A 1 mcg = 3.3 Units Max level age &lt;10 = 1.7 micromols/L age &gt;10 = 2.9 micromols/L</th>
<th>Vitamin D 1 mcg = 40 Units Target 25-OH Vit D level = 75-150 nmol/L</th>
<th>Vitamin E mg = units (approximately)</th>
</tr>
</thead>
<tbody>
<tr>
<td>&lt;6 weeks</td>
<td>600 micrograms (2,000 units)</td>
<td>10 micrograms (400 Units)</td>
<td>50 mg</td>
</tr>
<tr>
<td>6 weeks to 6 months</td>
<td>1,200 micrograms (4,000 units)</td>
<td>10 micrograms (400 Units)</td>
<td>50 mg</td>
</tr>
<tr>
<td>6 months to 1 year</td>
<td>2,400 micrograms (8,000 units)</td>
<td>10 micrograms (400 Units)</td>
<td>50 mg</td>
</tr>
<tr>
<td>1 – 10 years</td>
<td>2,400 micrograms (8,000 units)</td>
<td>10 – 20 micrograms (400 – 800 Units)</td>
<td>100 mg</td>
</tr>
<tr>
<td>11+ and Adults’</td>
<td>2,400 micrograms (8,000 units) = usual dose Max dose 3,000 units (10,000 units)</td>
<td>20 -50 micrograms (800 – 2000 Units)</td>
<td>100 -200 mg</td>
</tr>
</tbody>
</table>

*Individual doses will be adjusted by the specialist at the annual review according to vitamin levels.*
If vitamin D levels are deficient then the hospital may give a short course of high dose oral cholecalciferol (e.g. 20,000 units – Dekristol 2 -3 doses a week for between 1 and 4 weeks depending on how low the levels were), before requesting that the GP continues the licensed supplement below. The vitamin A&D product has maximum doses imposed to ensure that the individual is not given too much vitamin A.

Not all CF patients take Vitamin K. The recommended dose of vitamin K daily is not clear and is the subject of research. Currently, CF patients only take 10mg menadiol daily if they have some signs of liver involvement seen with abnormal LFTs/ clotting or changes on liver ultrasound at the annual review.

**Recommended supplementation**

1) **A&D….one of:**
   - Vitamins A & D BPC 1973
     - each capsule contains 4,000 Units vitamin A and 400 units vitamin D usual dose 2 daily (usual maximum dose 4 daily) **OR**
   - Dalivit 1.2ml supplies 3000 mcg of vitamin A, 20 mcg (800 units) of vitamin D **OR**
   - Calceos is given only if calcium intake is low and in older children/adults
     (1 tablet contains Calcium with 400 units Vitamin D)

2) **Vitamin E** suspension 100mg/ml

3) **Vitamin K** -
   - menadiol 10mg tablets. 1 daily is the usual dose.

***We do not recommend either Ketovite or Forceval***

**Is AquADEKs Solution Cost Effective?**


AquADEKs solution (60ml) is marketed in the USA and can be imported by Pharmarama (Tel 0208 238 6770) for use in the UK. It is prescribable but is an unlicensed product so classified as a “special” in the community. AquADEKs contains Vitamin A,D, E and K & is easy to take. GPs should put Pharmarama & their Tel no on prescriptions in the directions to facilitate community ordering.

AquADEKs solution contains
- Vitamin A 5751 units/ml
- Vitamin D 400 units/ml
- Vitamin E 50 units/ml
- Vitamin K 400 micrograms/ml

The usual daily dose is 1 ml daily if <1 year or 2ml daily for older children.

It is a useful and recommended source of vitamins (including vitamin K) for CF patients who are unable to swallow tablets to achieve the required level of supplementation.

Where patients are able to swallow capsules we recommend switching them over to a combination of Vitamin A&D capsules, Vitamin E suspension and, if needed, menadiol tablets as these preparations are all licensed.

**The AquADEKs softgel and chewable tablet are non-formulary**

**Note** - all fat soluble vitamins will be absorbed better if taken with a fatty food or drink such as milk.