11. Drug Formulary

11. Drug Formulary

11.1 DRUGS FOR THE RESPIRATORY TRACT

In CF, doses of antibiotics are usually given at a higher dose and for a longer period than in non-CF children, for reasons of pharmacokinetic differences as well as the presence of underlying lung disease. See section 6.2a for antibiotic prescribing policies.

NOTE: **od = once daily; bd = twice daily; tds = 3 times daily; qds = 4 times daily**

11.1a ORAL ANTIBIOTICS - PROPHYLACTIC DOSES

Azithromycin	Oral	<15kg: 10mg/kg od 3/week 15-40 kg: 250mg od 3/week >40kg: 500mg od 3/week NOTE dose differs from acute treatment	***Consultant decision*.** *Potential* for hepato- and ototoxicity but usually *very* well tolerated. Can cause tooth and tongue discolouration. **Avoid** long term concurrent use with erythromycin Can prolong QT interval. ECG before starting. For anti-inflammatory effect see section 11.1f other respiratory treatments.

Co-amoxiclav **400/57** (Augmentin duo)	Oral Susp	2 months – 2 yrs: 0.15 ml/kg bd; 2-6 yrs: 2.5 ml bd 7-12 yrs: 5 ml bd	Use only if regularly grows *H influenzae*. May discolour teeth. We do not use for Staph prophylaxis. **Clean teeth after dose**
Co-amoxiclav **125/31**	Oral Susp	<1 yr: 0.25ml/kg (max 5ml) bd	
Co-amoxiclav **250/62**	Oral Susp	1-<6 yrs: 2.5ml bd 6-12 yrs: 5ml bd	
Co-amoxiclav **250/125**	Oral tabs	>6 yrs: 1x (375 mg) tab bd	
Flucloxacillin	Oral	125mg bd (This dose is for prophylaxis in those < 3 years, and those in CF START study) Older children: 25 mg/kg bd (usual max 1 gm bd)	Give 1 hour BEFORE meals or on an empty stomach. Liquid tastes awful – different brands may be tolerated better than others. If *S aureus* a troublesome, regular problem can use up to 2 g bd - ***Consultant decision*.**

11.1b ORAL ANTIBIOTICS – TREATMENT DOSES

See section 6.2a for antibiotic prescribing policies. Decision depends on:

- Current clinical state.
- Current and past organisms and their antibiotic sensitivities.
- Past history of individual.
- Known 'allergies' or intolerance.

Azithromycin	Oral	10 mg/kg od max 500 mg NOTE dose differs from prophylactic and long term use Usually 10 days.	For *S aureus, H influenzae* and *mycoplasma* For anti-inflammatory effect see section 11.1f. other respiratory treatments. Ten days gives about 1 month's coverage. *Potential* for hepato- and ototoxicity but usually *very* well tolerated. Can cause tooth and tongue discolouration. Can prolong QT interval
Chloram-phenicol	Oral	>1 month: 12.5 mg/kg qds. Occasionally use 25 mg/kg qds (Max 4 gms/day). 2-3 weeks course	Consider for *S maltophilia, P aeruginosa, B cepacia, S aureus*. Needs full blood count at day 21 if course longer than 3 weeks. Very expensive (£450 - £1700 per two week course) Preferably round dose to the nearest whole capsule. Capsules can be opened, and the contents mixed with water or orange juice and given immediately. Levels should be considered in all patients receiving higher doses of 25mg/kg qds, children < 4 years old and patients with hepatic impairment. Aim trough level of <10 mg/L and 2 hour post dose level 10-25 mg/L after at least 1 day of therapy.

Ciprofloxacin	Oral	<1 month: 15 mg/kg bd ≥1month: 20 mg/kg bd (max 750mg) bd. Care should be taken if previously used within previous 3 months because of risks of resistance. **3 weeks for 1st isolation.** ***Consultant decision*** to exceed this length. Usually 2 weeks for exacerbations.	**First line** oral antipseudomonal agent. Photosensitising so warn patient re sunlight. High strength sunblock should be used in summer or on holidays for 4 weeks after course finished. Milk **will** reduce absorption. Avoid milk for at least 30 mins before and after taking ciprofloxacin. Also used for NTM treatment – ***consultant decision.*** See section 6.2a 6 VII. Can prolong QT interval. Joint pains occasionally – risk of tendonitis and tendon rupture – consider withdrawing treatment Parent/carer should be advised to stop ciprofloxacin and contact their doctor if they experience: - Tendon pain, swelling or rupture (can arise within 1 hour of starting treatment or up to 6 months after stopping) - Pain in joints or swelling in shoulder, arms or legs - Abnormal pain or sensations (*i.e*. tingling) esp. in legs or arms - Severe tiredness, depressed mood, anxiety, or problems with memory or severe problems sleeping - Change to vision, taste, smell or hearing
Clarithromycin	Oral	<8 kg: 7.5mg/kg bd 8 – 11kg: 62.5 mg bd 12 – 19kg: 125 mg bd 20 – 29kg: 187.5 mg bd 30 – 40kg: 250 mg bd (if >12 years old can increase to 500mg bd if necessary) 2-4 weeks	Cheaper alternative to azithromycin. Can cause tooth, tongue & urine discolouration. Part of NTM protocol. Care needed as interacts with some drugs *e.g.* itraconazole, rifabutin – check BNFc Can prolong QT interval

Clofazamine	Oral	<30kg: 50mg od 30kg and above: 100mg od Max dose 100mg od US/ECFS recommend 1-2 mg/kg BTS 3-5 mg/kg Our dose is a compromise	*Consultant decision* – reserved for the treatment of NTM. Take with or just after food. May cause a discoloration of the skin from red to brownish-black, as well as red staining of sweat, sputum urine, faeces, tears and saliva. Patient/carer should be advised that skin discoloration, although reversible, may take several months or years to disappear after the stopping therapy. Advise patient/carer to seek medical advice if persistent abdominal symptoms develop (pain, diarrhoea, nausea, vomiting). Can prolong QT interval. Do ECG prior to start of therapy and 2 weeks after, then if adding other drugs that can cause QT prolongation.
Co-amoxiclav **400/57** (Augmentin-Duo)	Oral susp	2 months – 2 yrs: 0.3 ml/kg bd; 2-6 yrs: 5 ml bd 7-12 yrs: 10 ml bd 1 month	For *S aureus* and *H influenzae* Care with CF liver disease
Co-amoxiclav **250/62**	Oral Susp	1-<6 yrs: 5ml tds 6-12 yrs: 10ml tds 1 month	
Co-amoxiclav **500/125**	Oral tablets	>6 yrs: (625mg tabs) 1 tab TDS 1 month	Co-amoxiclav 625mg tabs are to be used in preference to 2 x 375mg tabs to reduce clavulanic acid intake.
Co-trimoxazole	Oral	6 weeks–5 months: 120 mg bd 6 months–5 years: 240 mg bd 6–11 years: 480 mg bd 12–18 years: 960 mg bd 1 month	Use mainly for *S maltophilia* & MRSA. Maintain adequate fluid intake Treatment should be stopped if blood disorders or rashes develop. Advise patient/carer to report all rashes, sore throats and fevers. Avoid in severe liver disease.

Doxycycline	Oral	8-11years: 4.4mg/kg (max 200mg) **once daily** on day 1 - then 2.2mg/kg (max 100mg) once daily thereafter (can increase to 4.4mg/kg; max 200mg daily if required) ≥12 years: 200 mg **once daily** on day 1 - then 100 mg once daily thereafter (can increase to 200 mg daily if required). 2-4 weeks (can be used long term)	Can be useful for *S maltophilia* and *B cepacia*, and MRSA ***Consultant decision*.** Patient should be ≥12 years (due to discoloration of growing teeth and bone). However, can be used in 8-11 year olds in severe infection with no adequate alternatives, once confirmed with dental professional all 'adult' teeth in place. Take standing or sitting upright with 200 ml water (to avoid oesophageal irritation). Photosensitivity (see ciprofloxacin).
Ethambutol	Oral	20mg/kg od	***Consultant decision*** – reserved for the treatment of NTM. Monitoring - Visual acuity & colour vision testing at baseline and if symptoms reported, advise patients to report visual changes if possible. However, in very young children unable to report symptoms suggest routine ophthalmological examinations every 6 months. Peripheral neuropathy.
Flucloxacillin	Oral	30-35 mg/kg TDS MAX 4 gms/day 1 month	Give 1 hour BEFORE meals or on an empty stomach. Liquid tastes awful – different brands may be tolerated better than others.

Fusidic acid	Oral	<1 yr: 15mg/kg tds 1-4 yrs: 250 mg tds (5 ml) 5-12 yrs: 500 mg tds (10 mls) > 12 yrs: 750 mg tds (15mls) **or** 500mg sodium fusidate tablets tds 2 weeks	Caution in CF liver disease. Liquid should be taken with or after food Should always be prescribed with additional anti-staphylococcal agent Higher dose of fusidic acid liquid needed as incomplete absorption compared to sodium fusidate tablets.
Linezolid	Oral	<12 yrs: 10mg/kg (max 600mg) **tds.** ≥12 yrs: 600 mg bd 2 weeks	Last line for *MRSA* or *S aureus* where patients have not responded to conventional agents *e.g.* high dose flucloxacillin, rifampicin, fusidic acid. Occasionally used for NTM, consider use of pyridoxine (B_6) to reduce risk of cytopenias. ***Consultant decision*.** Courses >28 days leads to risk of optic neuropathy so patients having 4 week or repeated courses should have ophthalmic exam before starting first course and every 2 months after. Aim for 2 week courses. Where possible patients should be warned to immediately report any visual changes, regardless of treatment duration. Monitor FBC weekly.
Minocycline	Oral	8 - 11 years: 2mg/kg (max 100mg) bd ≥12 yrs: 100mg bd 2 weeks	Can be useful for *S maltophilia, B cenocepacia* & resistant *P aeruginosa.*. ***Consultant decision*.** Patient should be ≥12 years (due to discoloration of growing teeth and bone). However, can be used in 8-11 year olds in severe infection with no adequate alternatives, once confirmed with dental professional all 'adult' teeth in place. Caution in CF liver disease. Take standing or sitting upright with plenty of water (see doxycycline).

Moxifloxacin	Oral	7.5 – 10mg/kg (max 400mg) od	***Consultant decision*** – reserved for the treatment of NTM. **Not** active against *P. aeruginosa* or MRSA Joint pains occasionally – risk of tendonitis and tendon rupture – consider withdrawing treatment Parent/carer should be advised to stop ciprofloxacin and contact their doctor if they experience: - Tendon pain or swelling - Pain in joints or swelling in shoulder, arms or legs - Abnormal pain or sensations (*i.e*. tingling) esp. in legs or arms - Severe tiredness, depressed mood, anxiety, or problems with memory or severe problems sleeping - Change to vision, taste, smell or hearing Can prolong QT interval. The manufacturer advises should not be used concurrently with other drugs that prolong the QT interval: risks and benefits must be considered if this is deemed necessary. Do ECG prior to start of therapy and 2 weeks after, then if adding other drugs that can cause QT prolongation. Caution in CF liver disease.
Rifampicin	Oral	*S aureus* treatment: 10 mg/kg (max 600mg) bd. NTM treatment: 10 - 20 mg/kg (max 600mg) **od.** 2 weeks	Second line for *S aureus*. Usually give with fusidic acid. Occasionally used for NTM. Give 30 – 60 minutes ***before*** food. ***Consultant decision*.** Caution in CF liver disease. Please note rifampicin interacts with many drugs (including ivacaftor, Orkambi, clarithromycin, itraconazole, voriconazole, posaconazole, chloramphenicol) so always check in BNFc and with pharmacist. Check re oral contraceptive interactions. Can cause red staining of urine, tears and saliva.

11.1c INHALED ANTIBIOTICS

See NHSE Clinical Commissioning Policy for inhaled therapy first published Dec 2014.
https://www.england.nhs.uk/commissioning/wp-content/uploads/sites/12/2015/01/a01-policy-inhld-thrpy-cf.pdf

Amikacin (from IV solution)	Nebulised	6-12 years: 250mg bd (add 2ml 0.9% saline to 1ml of 250mg/ml amikacin). >12 years: 500mg bd (add 1ml 0.9% saline to 2ml of 250mg/ml amikacin). If not tolerated, retry at a lower dose.	Usually for NTM. Can further dilute injection with 0.9% sodium chloride. Suitable for jet nebuliser but not e-flow rapid. Can't use I-neb. Avoid using ear bud headphones for increased risk of hearing problems.
Amphotericin (Fungizone)	Nebulised	<10 years: 5 mg bd >10 years: 10 mg bd The dose can be increased up to 0.5mg/kg (max 25mg) bd depending on clinical response and tolerability	For chronic aspergillus. ***Consultant decision***. No need to use expensive liposomal preparation unless cannot tolerate standard preparation which tastes awful. Only suitable for Jet nebuliser. Can't use e-flow or I-neb. Dilution: 50 mg in 10ml of water for injection. Withdraw required dose and further dilute with water to a minimum volume of 3ml for nebulisation. Use 1 vial per day; keep remaining solution in the fridge
Aztreonam Lysine (Cayston)	Nebulised	75 mg tds during alternate months Mix with 1ml 0.17% saline (diluent comes with drug). Licensed >6 years Not commissioned for continuous use (only for alternate month).	3rd line for chronic *P aeruginosa.* Doses should be taken at least 4 hours apart. Pre dose with bronchodilator ***Consultant decision.*** Colistin or tobramycin usually given during the intervening month Should ideally be stored 2-8ºC. but can be kept out of the fridge, but below 25ºC, for up to 28 days. Used via e-flow but must use special Altera handset which nebulises to dry.

Ceftazidime	Nebulised	1 gm bd Reconstitute 1 gram injection with 3ml water for injection	For *B cepacia*. Tastes awful. ***Consultant decision*** Only suitable for Jet nebuliser. Can't use e-flow rapid or I-neb.
Colomycin (Colistin)	Nebulised	<8 yrs: 1,000,000 Units bd >8 yrs: 2,000,000 Units bd Mix with 3ml 0.9% saline. 1,000,000 units = 1 megaunit (Mu) Colomycin - Nebulise in Jet nebuliser or e-flow rapid. **Not** I-neb.	1st line for chronic *P aeruginosa.* Bronchospasm can be reduced by i) pre-dose with bronchodilator. and ii) diluting with salbutamol For Promixin 500,000 unit doses: the reconstituted solution may be kept for up to 24 hours in the fridge.
Promixin (Colistin)	Nebulised via I-Neb	< 8 years: 500,000 units bd >8 years: 1,000,000 units bd Promixin - I-neb only. Use Grey I-neb Chamber.	
Colobreathe turbospin (Colistin)	Inhaled (dry powder inhaler)	1 capsule (1.66 MU) bd via Turbospin powder inhaler Licensed >6 years only	Doses should be inhaled as close as possible to 12 hours apart. Put fat end of capsule into inhaler first to minimise capsule shattering when capsule is pierced.
Meropenem (from IV solution)	Nebulised	6-12 years: 125mg bd >12 years: 250mg bd	Usually for NTM. Also used for *B cepacia* chronic therapy. Reconstitute 500mg vial with 10ml 0.9% sodium chloride (keep remainder of vial in fridge for up to 18 hours). For a 250mg dose: use 5mls of reconstituted solution. For a 125mg dose: use 2.5mls of reconstituted solution and add 0.5mls of 0.9% saline Nebulise in Jet nebuliser not e-flow rapid.

Tobramycin – Bramitob TOBI Tymbrineb	Nebulised	300 mg bd during **ALTERNATE MONTHS** Licensed >6 years only Doses should be nebulised as close as possible to 12 hours apart and not less than 6 hours.	1st line for eradication of *P aeruginosa* given for 1 month. 2nd line for chronic *P aeruginosa.* ***Consultant decision*****.** In chronic *P aeruginosa* colistin will usually be given in the month off tobramycin. In our experience Bramitob® tends to be better tolerated in pre-school children and best given via a jet nebuliser. Use Jet nebuliser, E-flow or I-neb (lilac chamber); TOBI, Tymbrineb & Bramitob need to be nebulised twice per dose (2 fills per dose) if given via an I-neb (lilac chamber). After removal from refrigerator, TOBI or Tymbrineb pouches (intact or opened) may be stored at up to 25ºC for up to 28 days. Bramitob pouches (intact or opened) may be stored at up to 25ºC for up to 3 months. When switching between Bramitob and either TOBI or Tymbrineb a DRA is required as solutions are different concentrations; TOBI and Tymbrineb are interchangeable and don't require a DRA when switching.
Tobramcyin – TOBI Podhaler	Inhaled (dry powder inhaler)	112mg (4 x 28mg capsules) bd via podhaler during **ALTERNATE MONTHS** Licensed >6 years only	Doses should be inhaled as close as possible to 12 hours apart and not less than 6 hours.
Vancomycin	Nebulised	4mg/kg (maximum 200mg) qds for 5 days for eradication May be used bd for chronic suppression. *Consultant decision*	MRSA Reconstitute according to manufacturer's instruction (take into account displacement volume). Draw up required dose and make up to a total of 4ml with sodium chloride 0.9%. Use jet nebuliser. Pre-dose with nebulised salbutamol.

11.1d INTRAVENOUS ANTIBIOTICS

See section 6.2a for antibiotic prescribing policies. Decision depends on:

- Current and past organisms and their antibiotic sensitivities.
- Past history of the individual patient.
- Known 'allergies' or intolerance.

NOTE

i) Two antipseudomonal antibiotics from different classes are ALWAYS given – consultants only for exceptions.
ii) Add in IV teicoplanin if *S aureus* has been grown in the past year.
iii) Preferred *blind* starting combination is meropenem (better Staph cover) or ceftazidime **plus** tobramycin (gentamicin is never used due to increased renal toxicity and less favourable MIC).
iv) Consent MUST be taken and oral N-acetylcysteine (NAC) prescribed for all patients receiving IV aminoglycosides.
v) Course length is **always** a minimum two weeks.
vi) Take care with first doses as unexpected, severe hypersensitivity does occur.
vii) Antibiotics can impair liver and renal function. Take care with drug dosing with underlying impairment – refer to BNFc or the pharmacy team for more information.

We RARELY use:

i) Imipenem - too many side effects and spectrum no different from meropenem.

ii) Piperacillin/tazobactam (Tazocin, piptazobactam) is rarely used because there is a high incidence of allergy.

CIVAS (Centralised Intravenous Additives Service)

CIVAS is outsourced to an external provider.

Since most patients come in for admission during the daytime, the dose for that night and the next morning is made up by the nurses in the usual way on the ward. Admissions from Friday daytime, Saturday & Sunday (and bank holidays) will receive drugs made up on the ward until evening of next midweek working day.

Amikacin	IV	30 mg/kg od (max 1.5g od)	Aminoglycoside Infuse over 30 mins. Levels at 23 hours after 1st dose (*i.e.* **before** 2nd dose) must be < 3mg/l. Repeat at least every 7 days. If level raised, OMIT next dose and re-measure, reduce dose by 20%. See section 6.2a Only use if resistant to tobramycin or gentamicin. Used for initiation of NTM treatment – *consultant decision* Audiology at baseline. Consent MUST be taken for use of aminoglycosides EVERY TIME GIVEN; and oral N-acetylcysteine (NAC) prescribed for all patients receiving IV aminoglycosides.
Aztreonam	IV	50 mg/kg tds (Max 8 gms per day).	Monobactam No gram-positive activity.
Cefoxitin	IV	50mg/kg tds (Max 12g /day).	Cephalosporin Can give as a slow bolus or infusion over 30 minutes. Reserved for treatment of NTM – ***consultant decision.*** NOT active against *P aeruginosa.*
Ceftazidime	IV	50 mg/kg tds (Max 9 gms /day).	Cephalosporin Unexpected hypersensitivity on first exposure.
Ceftazidime/ Avibactam	IV	<12 years: 50mg/kg ceftazidime/12.5mg/kg avibactam tds (Max 2000mg ceftazidime/ 500mg avibactam tds) ≥ 12 years: 2000mg ceftazidime/ 500mg avibactam tds	Cephalosporin + beta-lactamase inhibitor Infuse over 120 minutes. Reserved for resistant *P aeruginosa*, NTM or *B cenocepacia*, as part of a 2nd line treatment regimen, based on in vitro sensitivities – ***Consultant decision.***

Colistin	IV	20,000-25,000 units/kg **tds**. **Long term use at home:** Use above total daily dose divided into 2 doses *i.e.* (30,000-38,000 units/kg **bd**)	Polymyxin Slow infusion over 30 mins. Max concentration is 40,000 units/ml. Boluses (over at least 5 mins) can be used for **Portacaths** only – not PICC lines. <12 yrs: dilute to 90,000 units/ml. ≥12 yrs: dilute to 200,000 units/ml. Measure renal function once a week. Not a first line agent**. Avoid using with IV amphotericin** (renal toxicity)**.**
Co-trimoxazole	IV	>6 weeks old: 60 mg/kg BD (no upper dose limit)	Useful for *A xylosoxidans & S maltophilia* ***Consultant decision*** Infuse over 60-90 minutes. Maintain adequate fluid intake. Treatment should be stopped if blood disorders or rashes develop. Advise patient/carer to report all rashes, sore throats and fevers. Avoid in severe liver disease.
Fosfomycin	IV	1-11 months (up to 10kg): 100mg/kg TDS 1-11 yrs (10-39kg): 125mg/kg TDS (Max 400mg/kg daily) 12-17 yrs (>40kg): 8g TDS	Useful for patients with multi-resistant *Pseudomonas aeruginosa* and/or where allergies are an issue – *consultant decision.* Fosfomycin is associated with a high sodium load - monitor fluid balance during therapy & U&E's twice weekly. Infuse in glucose 5% at a maximum rate of 133mg/minute.

Linezolid	IV	<12 years: 10mg/kg tds (max 600mg tds) ≥12 years: 600mg bd	Oxazolidinone Infuse over 30 – 120 mins. Monitor FBC weekly. ***Consultant decision*** **Courses >28 days lead to risk of optic neuropathy so patients having alternate monthly Linezolid should have ophthalmic exam before starting first course and every 2 months after.** Where possible patients should be warned to immediately report any visual changes, regardless of treatment duration. Use oral route wherever possible. Otherwise convert to oral route as soon as clinically indicated. Last line for *MRSA* or *S aureus* where patients have not responded to conventional agents.
Meropenem	IV	20 – 40 mg/kg tds. (Max 2g tds)	Carbapenem Give slowly over 5 minutes. Headache common.
Piperacillin / Tazobactam	IV	>1 month: 90mg/kg qds (Max 4.5g qds)	Ureidopenicillin. Give slowly over 5 minutes. ***Consultant decision*.** Not used unless we are desperate due to rashes and hypersensitivity
Teicoplanin	IV	>2 months – 11 years: 10mg/kg 12 hourly for 3 doses (loading dose) followed 24 hours later by 10mg/kg od. ≥12 years: 6mg/kg 12 hourly for 3 doses (loading dose) followed 24 hours later by 6mg/kg od (no upper dose limit)	Glycopeptide Can give as a slow bolus or infusion over 30 minutes ***Consultant decision*** Therapeutic drug monitoring may be of value in severe infection, MRSA and unexpected therapeutic failure. Discuss need with microbiologist. Aim trough 15 -60mg/L (taken after at least 7 days). Note levels take 5 days to come back.
Temocillin	IV	25mg/kg bd (Max dose 2g bd)	Penicillin Slow bolus over 3 – 5 minutes ***Consultant decision***. 3rd line

Tigecycline	IV	8 – 11 years: 1.2mg/kg (max 50mg) bd ≥12 years: 50mg bd, reduced to 50mg od if not tolerated	Tetracycline Reserved for treatment of NTM. **C*onsultant decision*.** Infusion over 60 minutes. Nausea/vomiting a real problem. Use regular IV Ondansetron – ensure that patient receives anti-emetics **before** commencing treatment. Before using in children <12 years old, please confirm with dental professional all 'adult' teeth in place (due to discolouration of growing teeth/bone).
Tobramycin	IV	10mg/kg/day in ONE DOSE (Max 660mg/day) If previous course had raised trough level reduce dose by 20%. Note this dose is for CF patients only.	Aminoglycoside Infuse over 30 mins. Levels at 23 hours after 1st dose (*i.e.* **before** 2nd dose) must be <1 mg/l) Repeat at least every 7 days. If level raised, OMIT next dose and re-measure. See section 6.2a Usual dilution: 0.9% sodium chloride. **DO NOT PRESCRIBE THIS DOSE FOR NON-CF CHILDREN.** Consent MUST be taken for use of IV aminoglycosides EVERY TIME GIVEN; and oral N-acetylcysteine (NAC) prescribed for all patients receiving IV aminoglycosides.

11.1e ANTIFUNGAL ANTIBIOTICS

Itraconazole	Oral	1month – 12 yrs: 5 mg/kg twice daily (max 200mg bd) >12yrs 200 mg twice daily	1st line for treatment of aspergillus infection in children aged 8 and below. Poorly absorbed, use liquid, on empty stomach if possible. Capsules should be taken with acidic liquid *e.g.* **Coca-Cola** and **food**. Stop antacids if possible. Headaches seem commonest problem but in theory hepatotoxic. Adrenal suppression also been seen when combined with budesonide. Do liver function tests if taken for longer than 1 month or if known liver dysfunction. Levels should be monitored if efficacy is not observed, concerns about toxicity, or if an interacting drug is commenced. Pre-dose samples taken after at least 2 weeks on therapy. Aim: 0.5 - 2mg/L (parent molecule) and total (including active metabolite) of 1-4mg/l. Note itraconazole interacts with many drugs (including **ivacaftor, Orkambi** and **rifampicin)** so always check in BNFc. See section 6.3 for length of courses.
Terbinafine	Oral	10 – 19kg: 62.5mg od 20 – 39kg: 125mg od 40kg +: 250mg od	For use in combination with an azole antifungal for *Scedosporium apiospermum* & *Lomentospora prolificans.* Consultant decision. Monitor liver function tests every 8 weeks when given in combination with an azole.

<table>
<tr>
<td rowspan="2">Posaconazole</td>
<td>Oral Suspension</td>
<td><8 years:
8 mg/kg TDS *

>8 years:
400mg BD *

Monitor levels.

Monitor liver function tests monthly.</td>
<td rowspan="2">1st line for ABPA for all ages.

1st line for aspergillus infection for 8 years and above
(use itraconazole in younger children)

Consultant decision (not licensed in <18 years old).

The tablet and oral suspension are not to be used interchangeably due to the differences in the dosing of each formulation. Tablets should be used preferentially as in our experience more consistent levels are obtained.

* Suspension should be taken immediately following a meal (preferably fatty meal) to enhance absorption. If this is not possible, may need to split the total daily dose into QDS dosing. Splitting the total daily dose into TDS or QDS dosing should also be considered if levels are <1mg/L.

Levels when using suspension reduced by ranitidine and proton pump inhibitors, which should be stopped if possible.

Tablets can be taken with or without a meal.

Levels should be monitored on initiation, on amendment of dosage, if an interacting drug is commenced or efficacy is not observed. Pre-dose samples (trough) taken after at least 1 week on therapy. Aim: 1 - 5mg/L For levels >5mg/L review dose with consultant and pharmacist.

Note posaconazole interacts with many drugs (including ivacaftor, Orkambi and rifampicin) so always check in BNFc and with a pharmacist.

See section 6.3 for length of courses.</td>
</tr>
<tr>
<td>Oral tablets</td>
<td>>8 years:
300mg BD on day 1, then 300mg OD thereafter

Monitor levels.

Monitor liver function tests monthly.</td>
</tr>
</table>

Voriconazole	Oral	**2 – 11 years:** 9mg/kg (max 350mg) bd (Liquid preferred) **12 - 14 years:** **<50kg** 9mg/kg (max 350mg) bd **>50kg** 400mg bd for 2 doses then 200mg bd (max 300mg bd). **15 years +:** **<40kg:** 200mg bd for 2 doses then 100mg bd (max 150mg bd) **>40kg**: 400mg bd for 2 doses then 200mg bd (max 300mg bd).	May be used for ABPA (3rd line) where patients have not responded to or are intolerant of itraconazole or posaconazole. ***Consultant decision*.** Take on an empty stomach. **Highly photosensitising** so warn patient re sunlight. High strength sunblock should be used in summer or on holidays for 4 weeks after course finished. Refer to dermatologist if photosensitivity reaction occurs. Risk of squamous cell carcinoma of the skin has been reported in long term use in patients with photosensitivity and other risk factors. Adrenal suppression has been reported in patients also taking inhaled corticosteroids. Levels should be monitored on initiation, on amendment of dosage, if an interacting drug is commenced or efficacy is not observed. Pre-dose samples taken after at least 3 days on therapy. Aim: 1.3 - 5.7mg/L Note voriconazole interacts with many drugs (including **ivacaftor, Orkambi** and **rifampicin)** so always check in BNFc and with a pharmacist. Monitor liver function tests + U&E's weekly for first month then monthly thereafter. See section 6.3 for length of courses
Liposomal amphotericin (Ambisome)	IV	5 mg/kg od Start at 1 mg/kg once daily then increase to 5 mg/kg od over 3 days. Give test **dose** of 100 mcg/kg (max 1mg) over 10 mins. Observe for 30 mins then continue treatment.	For invasive or troublesome aspergillus. Check **renal/liver function and U&Es at least 3/week.** **Use with caution with other nephrotoxic antibiotics** *e.g.* aminoglycosides, colistin. We NEVER use the standard amphotericin preparation (Fungizone) for IV use. Always prescribe using brand name. ***Consultant decision***. Administer over 30 mins. Compatible with 5% glucose only. Flush pre- & post dose with 5% glucose.

Caspofungin	IV	<3 months: 25 mg/m^2 od 3months - 1yr: 50 mg/m^2 od >1 yr: 70 mg/m^2 (max 70mg) on day 1 then 50 mg/m^2 (max 70mg) od. This can be increased to 70 mg/m^2 (max 70mg) od if lower dose is tolerated but inadequate response	For invasive or troublesome aspergillosis. ***Consultant decision*.** Reduce dose in liver impairment (see BNFc). Infuse over 60 mins. Incompatible with glucose solutions.

11.1f OTHER RESPIRATORY TREATMENTS

Aminophylline	IV	**Load:** 5mg/kg (max 500mg) over at least 20 minutes, then – **IV infusion:** <12 years: 1mg/kg/hour >12 years: 0.5 – 0.7mg/kg/hour	***Consultant decision*** Do not use loading dose if already receiving oral theophylline or aminophylline. Measure levels 4- 6 hours after starting infusion, and daily thereafter. Do not exceed 20mg/l. Care needed as interacts with some drugs *e.g.* clarithromycin, erythromycin, fluconazole, ciprofloxacin – check BNFc
Azithromycin (see 11.1a & 11.1b for standard antibiotic doses)	Oral	<15kg: 10mg/kg od 3/week 15-40 kg: 250 mg od 3/week >40kg: 500 mg od 3/week Mon/Wed/Fri	Potential long-term treatment as anti-inflammatory. ***Consultant decision*** *Potential* for hepato- and ototoxicity but usually *very* well tolerated. Can cause tooth and tongue discolouration. **Avoid** long term concurrent use with erythromycin Can prolong QT interval. Do ECG before starting.
RhDNase (Dornase alpha) Homecare delivery	Nebulised	2.5 mg once daily	Discuss timing with physio – usually in afternoon, at least 30 mins pre-physiotherapy. See section 6.5 for more details of variation of timing. Occasionally use twice daily - ***consultant decision.*** Jet nebuliser, E-flow Rapid or I-neb (Green chamber if I-neb) RhDNase should be kept in the fridge. A single brief exposure to elevated temperatures up to 24 hours at up to 30°C. does not affect its stability.
Hypertonic saline 3 or 7%	Nebulised	4 mls up to twice a day Immediately before or during physiotherapy.	Pre-treat with bronchodilator. (see section 6.6). Jet nebuliser, E-flow Rapid or I-neb (Lilac chamber if I-neb NB 2 fills per dose)

Ivacaftor	Oral	6 months and above: 5kg - <7kg: 25mg granules bd ≥7 - <14kg: 50mg granules bd ≥14kg - <25kg: 75mg granules bd ≥25kg: 150mg tablet bd	For children 6 months (and 5 Kg) and above with one of the following gating mutations- G551D, G1244E, G1349D, G178R, G551S, S1251N, S1255P, S549N, S549R. R117H 5T or 7T (not 9T) – post pubertal children • Must have clinical phenotype of CF and evidence of abnormal CFTR function (clinical or physiological). • We would not prescribe it to those with CFSPID. • Must fulfil the other criteria outlined within the NHSE Commissioning medicines for children policy (appendix 15). - Liver function tests 3 monthly for 1st year then yearly (annual review). - Eye exams before starting then annually in <12 yr olds. - Sweat chloride before starting and 6-8 weeks after starting, at 6 months, then annually. - Stool elastase in 2-5 year olds pre- and 6 months after starting. Take with fat containing food. • Sachet of granules should be mixed with one teaspoon (~5 mL) of age-appropriate soft food or liquid e.g. puréed fruits, yogurt, milk or juice. Once mixed, should be consumed within one hour. • Tablets must be swallowed whole and should not be chewed, broken or dissolved. • Doses should be given approximately 12 hours apart. Avoid food containing grapefruit or Seville oranges. Always check for interactions when initiating treatment with Ivacaftor or whenever new medicines are prescribed. See section 6.9 for specific drug interactions and refer to the paediatric pharmacy team for information.

Mannitol	Inhaled	Initiation dose assessment: see details in Summary of Product Characteristics on www.medicines.org.uk Therapeutic dose regimen: 400mg (10 x 40mg capsules) bd via inhaler supplied Licensed for >18 years only	***Consultant decision*** Commissioned for use in post-pubertal children provided NICE & NHSE criteria fulfilled. (see section 6.7). Doses should be taken morning and evening with evening dose taken 2 – 3 hours before bedtime. Tolerance should be assessed via 'Bronchitol Initiation Dose Assessment (BIDA)'
N-acetylcysteine (NAC)	Oral	< 12 years: 300mg BD ≥12 years: 600mg BD 600mg effervescent tablets.	Given for the duration of the IV aminoglycoside course. For the prevention of ototoxicity in patients receiving IV aminoglycosides.
Orkambi (Ivacaftor & Lumacaftor)	Oral	2 – 5 years: <14kg: Lumacaftor 100 mg/ivacaftor 125 mg sachets 1 sachet bd >14kg: Lumacaftor **150** mg/ivacaftor **188** mg sachets 1 sachet bd 6 – 11 years: Lumacaftor **100** mg/ivacaftor 125 mg tablets 2 tabs bd 12 years +: Lumacaftor **200** mg/ivacaftor 125 mg tablets 2 tabs bd Patients with very severe disease will be admitted to hospital for initiation of Orkambi (to allow a min of 4 hours observation	For children 2 years and above who are Phe508del homozygous - Liver function tests 3 monthly for 1st year then yearly (annual review). - Eye exams before starting then annually for all children <18 years. - Sweat chloride before starting then 6-8 weeks after starting (not mandatory). - Blood pressure before starting then periodically in clinic. Take with fat containing food. • Sachet of granules should be mixed with one teaspoon (~5 mL) of age-appropriate soft food or liquid e.g. puréed fruits, yogurt, milk or juice. Once mixed, should be consumed within one hour. • Tablets must be swallowed whole and should not be chewed, broken or dissolved. • Doses should be given approximately 12 hours apart. Concomitant use of azole antifungals is **not** recommended due to Orkambi markedly reducing levels of these antifungals.

		after first dosing). These patients will start on **1 tablet bd (half the usual dose)** and be closely monitored before increasing to usual dose if tolerated.	Always check for interactions when initiating treatment with Orkambi or whenever new medicines are prescribed. See section 6.9 for specific drug interactions and refer to the paediatric pharmacy team for information.
Symkevi (Ivacaftor & Tezacaftor)	Oral	12 years +: Symkevi (Tezacaftor 100mg/ivacaftor 150mg) 1 tab morning **And** Ivacaftor 150mg 1 tab in the evening	For children 12 years and above who are Phe508del homozygous **or** Phe508del heterozygous and have one of the following mutations: P67L, R117C, L206W, R352Q, A455E, D579G, 711+3A→G, S945L, S977F, R1070W, D1152H, 2789+5G→A, 3272-26A→G, and 3849+10kbC→T. - Liver function tests 3 monthly for 1st year then yearly (annual review). - Eye exams before starting then annually for all children <18 years. - Sweat chloride before starting then 6-8 weeks after starting (not mandatory) Take with fat containing food. • Tablets must be swallowed whole and should not be chewed, broken or dissolved. • Doses should be given approximately 12 hours apart. Avoid food or drinks containing grapefruit or Seville oranges. Always check for interactions when initiating treatment with Symkevi or whenever new medicines are prescribed. See section 6.9 for specific drug interactions and refer to the paediatric pharmacy team for information.

11.2 DRUGS FOR THE GASTROINTESTINAL TRACT

11.2a Pancreatic Enzymes

- Get to know one preparation properly. This clinic uses **Creon Micro (for infants) or Creon 10,000** for all children except under exceptional circumstances. See section 7.2 on PERT.
- Both creon preparations are porcine (pig) origin.
- Dose for a child established on pancreatic enzymes is *approximately* 1 capsule per 3-5 grams of fat.
- In babies, start with ½ scoop per feed (average fat content of 150ml standard infant milk is 5g) mixed with small amount of expressed breast milk, infant formula or apple puree*, just before feeds and increase in half scoop steps (quarters are too fiddly). Do not put Creon granules into the bottle.
- Enzymes may not be chewed or *mixed into* food, do not mix into hot foods
  - Dose should not exceed 10,000 units/kg/day of lipase without considering why needed.

Creon Micro = 5,000 units of lipase per scoop
Pancrex V powder= 25,000 units of lipase per gram
Creon 10,000 = 10,000 units of lipase per capsule
Creon 25,000 = 25,000 units of lipase per capsule

***NOTE:** At RBH we use apple puree to provide enzymes from birth as the puree keeps the enterically coated enzyme spheres in a suspension. This ensures that the child takes in the entire dose and minimizes the chance of gum breakdown caused by trapped enterically coated spheres in the mouth. If apple is not available, other fruit purees may be used. If apple purees for enzyme administration are introduced from birth, they must be done so carefully as it contradicts the WHO and Department of Health recommendations on the age that solids should be introduced to infants.

Patient/Parent/Carer advice available at -
http://www.medicinesforchildren.org.uk/pancreatin-pancreatic-insufficiency

11.2b Fat soluble vitamins

Empirically, the aim is to have plasma levels of vitamins A and E at upper limit of normal range. Daily recommendations from the CF Trust Nutrition Working Party 2016 are:

Age	**Vitamin A** *1 mcg = 3.3 IU*	**Vitamin D** *1 mcg = 40 IU*	**Vitamin E**	**Vitamin K**
< 1 Year	<450 mcg (1500 IU)	10 – 50 mcg (400 - 2000IU)	40 – 80 IU	< 2 years: 0.3mg/kg/day
1 - 3 Years	450 - 3000 mcg (1500-10,000 IU)	10 - 125 mcg (400 – 5000 IU)	50 – 150 IU	2 – 7 years: 5mg/day
4 – 7 years			150 – 300 IU	
>8 years			150 – 500 IU	5 – 10mg/day

Preparations:

- **DEKAs Plus** and **Paravit-CF** are brands of all-in-one multivitamins designed for people with CF containing vitamins A, D, E and K. DEKAs Plus also contains a number of other vitamins and trace elements.

- Our first line all-in-one vitamin preparation is **DEKAs Plus** as it approved for use by the Advisory Committee on Borderline Substances (ACBS), so GPs are more likely to prescribe continuing supplies in the community.

- We offer DEKAs Plus to all newborn screened children (including those who are pancreatic sufficient). If children will not tolerate it, or if GPs are unable to continue supplies, then we will use Dalivit and vitamin E in infants.

- We will offer Paravit-CF to older children who do not tolerate DEKAs. Generally, though we use Paravit-CF for those with liver disease (defined as those on ursodeoxycholic acid).

- All patients will be supplied with a supply letter outlining information about the preparations for the GP and community pharmacist.

Vitamin	**DEKAs Plus Liquid (per ml)**	**DEKAs Plus Chewable tab**	**DEKAs Plus Softgel**	**Paravit - CF Liquid (per 0.125ml)**	**Paravit-CF Capsule**	**Dalivit (per 1.2ml)**
A	1,742mcg	5,505mcg	5,505mcg	757.5 mcg	1500mcg	3000mcg
D	750 IU	2000 IU	3000 IU	750 IU	1500 IU	400 IU
E	50 IU	100 IU	150 IU	75 IU	150 IU	Nil
K	0.5mg	1mg	1mg	2.5mg	5mg	Nil

- Abidec: not usually given due to low vitamin A content however may be a suitable alternative if Dalivit unavailable.

- One **vitamin A+D capsule BPC** contains – vitamin A 1200 mcg, vitamin D 10 mcg

- **Vita-E gel capsules**: 75 unit capsule ≈ 50 mg vitamin E
  400 unit capsule ≈ 268 mg vitamin E
  (Note that 200iu capsules no longer available from GPs)

Recommended dosing (empirical):

Birth to 12 months:

- Either **DEKAs** Plus Liquid 1ml od
- Or **Dalivit** 0.6 ml + **Vitamin E Liquid** 50 mg (0.5ml) od

1 to 4 years:

- Either **DEKAs** Plus Liquid 2ml od
- Or **Dalivit** 1.2 ml + **Vitamin E Liquid** 100 mg (1ml) od

5- 8 years:

- Either **DEKAs** Plus Liquid 2ml od or **DEKAs** Plus softgel or chewable tablet 1 od
- Or **Dalivit** 1.8 ml + **Vitamin E Liquid** 100 mg (1ml) od

9 years and above:

- Either **DEKAs** Plus 1-2 softgels or 1-2 chewable tablets od
- Or **Vitamin A&D capsules** 2-3 + **Vitamin E (Vita-E Gel 75iu/400iu Caps)** 150 - 400iu.

Paravit-CF

0-12 months	Liquid 0.125 mls od
1-4 yrs	Liquid 0.25 mls od
5-8 yrs	Liquid 0.25 mls od OR Capsule 1 od
9 yrs+	Capsules 1-2 od

Note: annual review blood levels may not reflect dosages prescribed as low levels may simply reflect poor adherence.

Vitamin D deficiency (see section 8.4)

Anyone with a vitamin D level below 50nmol/l should be treated.

Stoss therapy is the default therapy. It involves a single oral administration of the total treatment dose of vitamin D. An alternative is the whole dose as a single intramuscular injection but there is no reason to use this. This may need to be repeated, if poor compliance persists with maintenance dosing. However, the Sydney paper (Shepherd et al, JCF 2012) showed this regimen maintained vitamin D levels for a year.

Oral colecalciferol **single dose**:

- 1 - 12 months 150,000 units
- 1 - 12 years 300,000 units
- ≥12 years 500,000 units

Also available as 50,000 units in 1 ml oral ampoule (Invita D3).

The previous regimen can still be used if there are difficulties with prescribing high dose stoss therapy –

Oral colecalciferol for **3 months**:

- Infant 1 - 6 months 3000 units daily
- 6 months - 12 years 6000 units daily
- ≥ 12 years 6000 - 10,000 units daily
- Alternative for older children – colecalciferol 20,000 units 3 times a week; or colecalciferol 50,000 units once a week.

This can be as

- colecalciferol liquid 3000 units/ml.
- colecalciferol capsules or dispersible tablets 1000 units
- colecalciferol capsules 10,000 or 20,000 units
- colecalciferol liquid 50,000 unit/1ml

Vitamin K

We are now using **Paravit-CF** as the routine multivitamin for those with liver disease as it contains sufficient vitamin K that we do not have to prescribe separate menadiol or phytomenadione. This is given instead of DEKAs. See above for dosing.

If there are significant clotting abnormalities and extra vitamin K is required use~~:~~ **Menadiol phosphate (water soluble) or Phytomenadione (fat soluble)** tablets (menadiol preferred when available). Menadiol can be dispersed in water if necessary.

Dose for 6 years & above: 10 mg od.

11.2 c 'Antacids'

If enzyme dose high and compliance and diet etc. have been assessed, then consider:

- **Oral ranitidine**: <1 month: 2 mg/kg tds (max 3 mg/kg tds)
  1 – 6 months: 1 mg/kg tds (max 3 mg/kg tds)
  >6 months: 2-4 mg/kg bd (max 150 mg bd)
  – small risk of headache.

- **Oral omeprazole**:

	Once daily dose	Maximum daily dose
<2.5kg	0.7 - 1.4mg/kg	3 mg/kg/day
2.5 – 7kg	5mg	3mg/kg/day (max 10mg/day)
7-15 kg	10mg	20 mg daily
>15kg	20mg	40 mg daily

- Doses may be divided and given twice daily if required.
- If using dispersible 'MUPS' tablets:
  - Round to nearest 5mg (half of a tablet).
  - Tablet can be cut in half but should not be crushed or chewed. Do not try to give a fraction of a tablet by dispersing it – it does not disperse evenly!
  - Allow tablet (or portion of) to dissolve on the tongue or disperse in water/juice/yoghurt and give the whole amount.
- Alternatively, patients can open the capsule and swallow the contents with half a glass of water or after mixing the contents in a slightly acidic fluid *e.g.*, fruit juice or applesauce, or in non-carbonated water. If using this method, then doses should be rounded to nearest 10mg (whole capsule).

- For administration through an *enteral feeding tube*, oral liquid or the contents of a Losec capsule dispersed in 10 mL Sodium Bicarbonate 8.4% (1 mmol Na^+/mL).
- If unable to tolerate omeprazole – lansoprazole can be tried as an alternative – see BNFc for doses.

11.2d Gastro-oesophageal reflux

Very common in CF.

- **Oral omeprazole**:see above (11.2c) for doses

  OR

- **Oral ranitidine** see above (11.2c) for doses

Consider: **Infant gaviscon**, <4.5kg: 1 dose per feed (max 6 doses/day); >4.5kg: 2 doses per feed (max 12 doses/day).

Erythromycin dose for gastric stasis is: 3 mg/kg qds orally.
- Risk of cardiac adverse effects such as arrhythmias.
- A baseline ECG is recommended in patients concomitantly receiving drugs that increase QT prolongation.
- Subsequent 6 monthly ECG's should only be completed for patients at an apparent risk. (*e.g.* cardiovascular instability)

11.2e Distal Intestinal Obstruction Syndrome (DIOS)

Old name meconium ileus equivalent (MIE). See **section 7.7 for treatment protocol.** All therapies are osmotic in action therefore fluid support is CRUCIAL, if necessary, intravenously.

- **Oral Gastrografin**: <15 kg, 25 ml BD with 75 ml flavoured juice / water
  15-25 kg, 50 ml BD with 150 ml flavoured juice / water
  >25 kg, 100 ml BD with 200 ml flavoured juice / water

Use for up to 3 days if no response in first 24 hours (but not if symptoms worsen).

Do NOT give in the presence of bile stained vomiting or bowel obstruction.

- **Rectal Gastrografin**: Use same doses as oral.
  <5yrs: Dilute to 5 times its volume with water
  >5yrs: Dilute to 4 times the volume with water
  Requires IV line for IV fluids.

- **Oral N-acetylcysteine** - tastes like rotten eggs – The 200mg/ml injection can be given orally and should be mixed with water, orange juice, blackcurrant juice or cola to a concentration of 50mg/ml. Alternatively, 200mg sachets or 600mg tablets are available.

1month – 2 years	0.4 - 3g STAT
2 – 6 years	2 – 3g STAT
≥7 years	4 – 6g STAT

- **Polyethylene glycol** (**Klean-prep)**

  - **Do NOT give in the presence of bile stained vomiting.**
  - Solid food should not be given for at least 2 hours before starting treatment.
  - Add contents of 1 sachet to 1 litre water – can be flavoured with a clear fruit cordial. After reconstitution the solution can be kept in a refrigerator and discarded if unused after 24 hours.
  - Can be given orally or via NG tube (usually latter) and a single dose of domperidone 30 minutes before starting can increase gastric emptying.
  - Do not administer just before bedtime due to risk of aspiration.
  - Patients must be reviewed after 1st 4 hours.
  - If not passing essentially clear fluid per rectum then a further 4 hours treatment can be given.
  - Monitor for hypoglycaemia, which can occur with CF diabetics undergoing this regimen.

  - Start at 10ml/kg/hour for 30 mins then 20 ml/kg/hour for 30 mins.
  - If well tolerated rate can go up to 25 ml/kg/hour.
  - Maximum volume is 100 ml/kg or 4 litres (whichever is smaller) over 4 hours.
  - 10ml/kg/hour for 30 mins, increased to 20ml/kilogram/hour for 30 mins.
  - Then if tolerated to 25ml/kg/hr, max 100ml/kg or 4 litres (whichever is the smaller volume) over 4 hours.
  - Repeat 4 hour treatment if necessary.

- **Oral Picolax:**
  1 - 2 years: 0.25 sachet STAT
  2 – 3 years: 0.5 sachet STAT
  4 – 8 years: 1 sachet STAT
  9 years and over: 1 sachet STAT

Prevention of DIOS:

N-acetylcysteine (oral)

<2 years: 100 – 200mg tds
2 – 11 years: 200mg tds

≤12 years: 200 – 400mg tds

11.2f Constipation

Ensure fluid intake is adequate.

Lactulose

<1 year 2.5 ml bd
1-4years: 2.5 – 10ml bd
>5years: 5-20 ml bd
then adjust dose according to response.

Movicol

Chronic constipation, prevention of faecal impaction:

1 - 5 years: 1 sachet of Movicol **Paediatric** OD.
Adjust dose accordingly - maximum 4 sachets daily.

6 - 11 years: 2 sachets of Movicol **Paediatric** OD.
Adjust dose accordingly - maximum 4 sachets daily.

12 - 17 years: Initially 1 - 3 sachets of Movicol per day in divided doses for up to 2 weeks. Maintenance dose 1-2 sachets daily.

Mix contents of each Movicol **Paediatric** sachet in 1/4 of a glass (60-65ml) water and each Movicol sachet in 1/2 of a glass (125ml) water

11.2g Liver disease

Ursodeoxycholic acid: 10 – 15mg/kg bd oral.
There is no advised maximum dose, but our adult unit would go up to 500 mg 2-3 times daily.
Commonest side effect is diarrhoea (rare though), in which case, reduce dose. Last dose should be taken in late evening.

Vitamin K – given as **Paravit-CF** (see 11.2b).

11.2h Anti-emetics

A pre-emptive and stepwise approach is necessary in preventing the impact of emetogenic medicines such as tigecycline:

1st line: Commence regular Ondansetron IV 100mcg/kg (max 4mg if < 12 years; 8mg if > 12 years) tds **before** starting treatment. Be aware it can be constipating, so if DIOS an issue ensure well hydrated and consider laxatives.

2nd line: If not controlled on above, consider adding regular domperidone PO 0.25mg/kg (max 10mg) tds

Note – both ondansetron and domperidone can prolong QT interval – baseline ECG should be carried out in patients concomitantly receiving drugs that increase QT prolongation.

3rd line: If nausea not controlled despite adequate doses of ondansetron and domperidone, or if above contraindicated, then add aprepitant 3mg/kg (max 125mg) on day 1 then 2mg/kg (max 80mg) on days 2 & 3. Stop after day 3 and reassess symptoms.

11.3 Home delivery of medicines

NHS England Prescribed Specialised Services Commissioning Intentions (2014) has stated that responsibility for the ongoing prescription of high cost inhaled medicines (dornase, tobramycin, colistin, aztreonam, mannitol) for cystic fibrosis should defer to the Hospital Trust. A homecare delivery service to supply these medicines directly to patients is already in use, as responsibility for any **new** prescriptions of these medicines (aside from those already prescribed by GPs) transferred to the Trust in 2014. However, the repatriation of those medicines currently being prescribed by GPs, to the trust is still being discussed.

This homecare service enables these medicines that are not able to be prescribed by the patient's GP, to be prescribed by the CF team at RBH, and then delivered directly to the patient at home by the hospital's chosen homecare provider, for as long as is required. The default for prescribing and supply of **all other** CF medicines except the ones listed above should be from the GP. Note if we prescribe from our pharmacy there is an extra 20% cost for VAT; if home prescribed then there is no VAT payable, so this is the preferred option.

If homecare is required then please contact a member of the paediatric pharmacy team as soon as possible (Bleeps 7403/7410/7425/7428 or ext. 84375; paedpharmacy@rbht.nhs.uk; rbh-tr.paediatricpharmacy@nhs.net) who will then advise on the process to be followed. The paediatric pharmacy team should also be informed if there are ANY CHANGES to patient medicines that are supplied via homecare *i.e*. dose changes or discontinuations. Where possible copy the paediatric pharmacist into correspondence detailing such changes.