

## Bristol Paediatric Cystic Fibrosis Formulary

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| Acetylcysteine  | Oral      | <2 yrs: 0.4-3 grams single dose<br>2-7 yrs: 2-3 grams single dose<br>>7 yrs: 4-6 grams single dose  | For treatment of DIOS. Use oral granules.  |
| Amikacin<br>( <i>Aminoglycoside</i> )                           | IV        | 30 mg/kg od (max 1.5 grams od)<br>Infusion over 20-30 mins.   | Level at 22-24 hours after 1 <sup>st</sup> dose must be <3 mg/l. Repeat day 9.   |
| Amoxicillin   | Oral      | <1 yr: 125 mg tds<br>1-7 yrs: 250 mg tds<br>>7 yrs: 500 mg tds  | Give at least 2 weeks.   |
| Amphotericin  | Nebulised | <10 yrs: 5 mg bd<br>>10 yrs: 10 mg bd   |  |
| Amphotericin<br>(liposomal)                                     | IV        | Start at 1 mg/kg od & increase<br>in daily steps of 1mg/kg to<br>3mg/kg od  | Give test dose of 100 mcg/kg (max<br>1 mg) over 10-15 mins. Check renal<br>function 3 times per week; be<br>careful if on other nephrotoxics.                          |
| Azithromycin<br><i>Treatment course</i>                         | Oral      | 10 mg/kg od (max 500 mg od)   | Usually give for 7-10 days.  |
| Azithromycin<br><i>Regular treatment</i>                        | Oral      | 15-40 kg: 250 mg od<br>>40kg: 500 mg od   | Alternative is 3 times per week.   |
| Aztreonam<br>( <i>Beta-lactam</i> )                             | IV        | 50 mg/kg tds<br>Can use qds (max 2 grams qds)   |  |
| Cephadrine  | Oral      | <12 yrs: 25 mg/kg bd<br>>12 yrs: 1 gram bd  | Give at least 2 weeks.   |
| Cefixime  | Oral      | <10 yrs: 8 mg/kg od (max<br>400mg)<br>>10 yrs: 200-400 mg od  | Give at least 2 weeks.   |
| Ceftazidime<br>( <i>Cephalosporin</i> )                         | IV        | 50 mg/kg tds (max 3 grams tds)  | Alternative is 75 mg/kg bd (max 4.5<br>grams bd).  |
| Ceftazidime   | Nebulised | Up to 1 gram bd   |  |
| Cefuroxime<br>( <i>Cephalosporin</i> )                          | IV        | 50 mg/kg tds<br>(max 1.5 grams tds)   |  |
| Chloramphenicol   | Oral      | 12.5 mg/kg qds<br>(max 1 gram qds)  | Smallest dose is 250 mg (capsule).<br>Needs microbiology approval.<br>Monitor FBC weekly.  |
| Ciprofloxacin   | Oral      | <5 yrs: 15 mg/kg bd<br>>5 yrs: 20 mg/kg bd (max 750<br>mg bd)   | Warn of photosensitivity – need to<br>take precautions.  |
| Clarithromycin  | Oral      | <8 kg: 7.5 mg/kg bd<br>8-11 kg: 62.5 mg bd<br>12-19 kg: 125 mg bd<br>20-29 kg: 187.5 mg bd<br>30-40 kg: 250 mg bd<br>>12 yrs: 250-500 mg bd |  |
| Clindamycin   | Oral      | 5-7 mg/kg (max 600 mg) qds  | Capsules are 250 mg.   |
| Co-amoxiclav<br>(Augmentin®)<br><i>Treatment course</i>         | Oral      | <6 yrs: 0.5 ml/kg of 125/31 tds<br>6-12 yrs: 0.3ml/kg up to 10 ml<br>of 250/62 tds<br>12-18 yrs: 1 tablet 500/125 tds                       | Give at least 2 weeks.   |
| Co-amoxiclav<br>(Augmentin®)<br><i>Prophylactic<br/>therapy</i> | Oral      | <6 yrs: 0.25 ml/kg of 125/31 bd<br>6-12 yrs: 5 mL of 250/62 bd<br>>12 yrs: Augmentin oral tabs<br>(250/125) 1 tab bd                        | Can use if flucloxacillin not<br>tolerated or if regular <i>H. influenzae</i> .<br>May discolour teeth. Septrin is<br>alternative. Augmentin-Duo® may<br>be an option. |

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| Colistin<br>(Colomycin®)                      | Nebulised | <1yr: 500,000 units bd<br>1-7 yrs: 1,000,000 units bd<br>>8 yrs: 2,000,000 units bd   | Can also use od. Be careful of bronchospasm; give first dose in hospital. Give after physio.   |
| Colistin<br>(Polymyxin)                       | IV        | 25,000 units/kg tds (max 2,000,000 units tds)   | Need to monitor renal function at least once per week. Avoid using with IV aminoglycosides and amphotericin.   |
| Co-trimoxazole<br>(Septrin®)                  | Oral      | <6 months: 120 mg bd<br>6 months – 6 years: 240 mg bd<br>6 -12 yrs: 480 mg bd<br>>12 yrs: 960 mg bd   |  |
| Dalivit                                       | Oral      | 0.6 ml od   | Dose guided by serum levels of vitamin A and D.  |
| rDNase<br>(Pulmozyme®)                        | Nebulised | 2.5 mg od   | At least 1 hour pre-physio. Occasionally use twice daily. Can consider using alternate days after 3 months.  |
| Domperidone                                   | Oral      | <12 yrs: 0.2-0.4 mg/kg (max 20mg) bd, tds, or qds<br>>12 yrs: 10-20 mg bd, tds or qds   |  |
| Doxycycline                                   | Oral      | >12 yrs: 200 mg od on 1 <sup>st</sup> day; then 100 mg od (can increase to 200mg od)  | Over 12 yrs only. Warn of possible photosensitivity. Take standing or sitting upright with water.  |
| Erythromycin                                  | Oral      | <2 yrs: 250 mg bd<br>2-8 yrs: 500 mg bd<br>>8 yrs: 500 mg - 1 gram bd   |  |
| Flucloxacillin<br><i>Treatment course</i>     | Oral      | 25 mg/kg qds (max 1 gram qds)   | Can use total daily dose in 2 or 3 doses to aid adherence.   |
| Flucloxacillin<br><i>Prophylactic therapy</i> | Oral      | 25 mg/kg bd   |  |
| Forceval Junior                               | Oral      | >5 yrs: 2 tablets od  | May be given as an adjunct to A & D capsules if vit D levels are low.  |
| Forceval                                      | Oral      | 12-18 yrs: 1 capsule od   | As for Forceval Junior.  |
| Gastrograffin                                 | Oral      | <10 kg: 25 ml with 100 ml flavoured juice / water<br>10-25 kg: 50 ml with 200 ml flavoured juice / water<br>>25 kg: 100 ml with 400ml flavoured juice / water | Highly osmotic; need careful hydration. Do not give if there is bile stained vomiting.   |
| Gentamicin<br>(nebulised)                     | Nebulised | <2 yrs: 40 mg bd<br>2-8 yrs: 80 mg bd<br>>8 yrs: 160 mg bd  | Be careful of bronchospasm; give first dose in hospital. Give after physio.  |
| Gentamicin<br>(Aminoglycoside)                | IV        | 10 mg/kg od (max 600mg od)<br>Infusion over 20-30 mins.   | Level taken 22-24 hours after 1 <sup>st</sup> dose must be <1 mg/l. Repeat on day 9.   |
| Hypertonic saline                             | Nebulised | 2-4 mls of 7% solution  | Up to twice per day 30 mins before physio.   |
| Itraconazole                                  | Oral      | 2.5 mg/kg bd<br>(Can also use 5 mg/kg od)   | Take with cola or other acidic liquid. Stop antacids if possible, or give 1 hour later. Consider monitoring LFTs. Consider monitoring levels (ideal trough serum level is 0.5-1 mg/l). |

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| Lactulose  | Oral      | <1 yr: 2.5ml bd<br>1-5 yrs: 5 ml bd<br>5-10 yrs: 10 ml bd<br>>10 yrs: 15ml bd             | Adjust according to response.   |
| Linezolid  | Oral      | <12 yrs:10 mg/kg tds (max 600 mg tds)<br>>12 yrs: 600 mg bd                               | Needs microbiology approval. Last line antibiotic against MRSA or <i>S aureus</i> . Monitor FBC weekly.   |
| Meropenem<br>( <i>Carbapenem</i> )                                     | IV        | 40 mg/kg tds (max 2 grams tds)  |   |
| Ketovite liquid  | Oral      | 5 mls od  | Rarely given as vit A content is low. May be useful if serum vit A is high.                               |
| Ranitidine   | Oral      | <6 months: 1 mg/kg tds<br>>6 months: 2-4 mg/kg bd (max 150mg bd)                          |   |
| Rifampicin   | Oral      | <1 yr: 5-10 mg/kg bd<br>>1 yr: 10 mg/kg bd (max 600 mg bd)                                |   |
| Sodium fusidate  | Oral      | <1 yr: 15 mg/kg tds<br>1-5 yrs: 250 mg tds<br>5-12 yrs: 500 mg tds<br>>12 yrs: 750 mg tds |   |
| Tazocin®<br>( <i>ureidopenicillin &amp; beta-lactamase inhibitor</i> ) | IV        | 90 mg/kg qds (max 4.5 grams qds)  | Piperacillin / tazobactam.  |
| Teicoplanin<br>( <i>Glycopeptide</i> )                                 | IV        | 10 mg/kg (max 400 mg) every 12 hours for 3 doses; then 10 mg/kg od (max 400 mg od)        |   |
| Timentin®<br>( <i>Carboxypenicillin</i> )                              | IV        | 80 mg/kg qds (max 3.2 grams qds)  | Ticarcillin / clavulanic acid.  |
| Tobramycin<br>( <i>Aminoglycoside</i> )                                | IV        | 10 mg/kg od (max 600mg od)<br>Infusion over 20-30 mins.                                   | Level taken 22-24 hours after 1 <sup>st</sup> dose must be <1 mg/l. Repeat on day 9.                      |
| Tobramycin<br>(IV solution)  | Nebulised | <2 yrs: 40 mg bd<br>2-8 yrs: 80 mg bd<br>>8 yrs: 160 mg bd                                | Be careful of bronchospasm; give first dose in hospital. Give after physio.                               |
| Tobi®  | Nebulised | 300mg bd  | Use on alternate months.  |
| Trimethoprim<br>( <i>Prophylaxis</i> )                                 | Oral      | <12 yrs: 2 mg/kg nocte (max 100 mg)<br>>12 yrs: 100mg nocte                               |   |
| Urokinase  | IV        | 5000 – 10 000 units in 0.9% saline (2mls)   | For occluded ports. Leave for 2-4 hours and then aspirate the lysate; then flush with heparinised saline. |
| Ursodeoxycholic acid   | Oral      | 10 mg/kg bd   |   |
| Vancomycin<br>( <i>Glycopeptide</i> )                                  | IV        | 15 mg/kg tds (max 660 mg tds)   | Trough level should be 5-10mg/l.  |
| Vitamin A+D  | Oral      | 1-2 capsules od   | These became unavailable from November 2007   |
| Vitamin BPC capsule  | Oral      | Usually start at 2 capsules od  |   |
| Vitamin E suspension   | Oral      | Infants: 0.5 ml od<br>>1 yr: 0.5 - 1ml  | May give up to 2ml od depending on serum levels. 1ml = 100mg.   |

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| Vitamin E tablets (Ephynal) 100mg | Oral | >1 yr: 100 mg od (1 tablet)  | May increase to 200mg; dose guided by serum levels.   |
| Vitamin K (Menadiol)              | Oral | 10 mg od   | Use in liver disease. Supplementation may also be important in bone mineralisation in other children with CF. |
| Voriconazole                      | Oral | <12 yrs: 6 mg/kg bd (max 200 mg) for 1 day, then 4 mg/kg bd (max 100 mg)<br>>12 yrs (<40 kg): 200 mg bd for 1 day, then 100 mg bd<br>>12 yrs: (>40 kg) 400 mg bd for 1 day, then 200 mg bd | Warn of photosensitivity and temporary visual problems<br>Monitor LFTs.                                       |
| Zinc (Solvazinc)                  | Oral | ½ tablet od (22.5 mg)  | Given if serum levels are low. Additional dietary advice given to try to improve intake.                      |