



A lifetime of specialist care

Other respiratory treatments

Aminophylline	IV	<p>Load: 5mg/kg (max 500mg) over at least 20 minutes, then –</p> <p>IV infusion: <12 years: 1mg/kg/hour</p> <p>>12 years: 0.5 – 0.7mg/kg/hour</p>	Consultant decision	<p>Do not use loading dose if already receiving oral theophylline or aminophylline.</p> <p>Measure levels 4- 6 hours after starting infusion, and daily thereafter. Do not exceed 20mg/l.</p> <p>Care needed as interacts with some drugs e.g. clarithromycin, erythromycin, fluconazole, ciprofloxacin – check BNFC</p>
Azithromycin (see 11.1a & 11.1b for standard antibiotic doses)	Oral	<p><15kg: 10mg/kg od 3/week</p> <p>15-40 kg: 250 mg od 3/week</p> <p>>40kg: 500 mg od 3/week</p> <p>Mon/Wed/Fri</p>	<p>Potential long-term treatment as anti- inflammatory.</p> <p>Consultant decision</p>	<p><i>Potential</i> for hepato- and ototoxicity but usually very well tolerated.</p> <p>Avoid long term concurrent use with erythromycin</p>
DNase (Dornase alpha) Homecare delivery	Nebulised	<p>2.5 mg once daily</p> <p>Consideration of alternate day after 6 months if well or treatment burden an issue.</p>	<p>In afternoon, usually at least 30 mins pre- physiotherapy.</p> <p>See section 6.4 for more details of variation of timing.</p>	<p>Occasionally use twice daily – consultant decision.</p>
Hypertonic saline 3 or 7%	Nebulised	<p>4 mls up to twice a day</p> <p>Immediately before or during physiotherapy.</p>	<p>Pre-treat with bronchodilator. (see section 6.5).</p>	
Ivacaftor	Oral	<p>2 years and above : <14kg: 50mg granules bd</p>	<p>For children with one of the following gating</p>	<p>Doses should be taken morning and evening with</p>

		<p>≥14kg - <25kg: 75mg granules bd</p> <p>≥25kg: 150mg tablet bd</p>	<p>mutations- G551D, G1244E, G1349D, G178R, G551S, S1251N, S1255P, S549N, S549R.</p> <p>-Liver function tests 3 monthly for 1st year then yearly (annual review). -Sweat chloride 6- 8 weeks after starting, at 6 months, then annually. -Eye exams before starting then annually in <12 yr olds. -Stool elastase in 2-5 yr olds pre- and 6 months after starting.</p>	<p>evening dose taken 2 – 3 hours before bedtime.</p>
Mannitol	Inhaled	<p>Initiation dose assessment: see details in Summary of Product Characteristics on www.medicines.org.uk</p> <p>Therapeutic dose regimen: 400mg (10 x 40mg capsules) bd via inhaler supplied</p> <p>Licensed for >18 years only</p>	<p>Consultant decision</p> <p>Not currently commissioned for use in children, and funding should be sought before initiation of treatment.</p>	<p>Doses should be taken morning and evening with evening dose taken 2 – 3 hours before bedtime.</p>
N-acetylcysteine (NAC)	Oral	<p>< 12 years: 300mg BD</p> <p>≥12 years: 600mg BD</p>	<p>Given for the duration of the IV aminoglycoside course. For the prevention of ototoxicity in patients receiving IV aminoglycosides as part of NTM treatment and 3- monthly IV's only.</p>	<p>600mg tablets (unlicensed brand ACC® from Hexal) are scored and halve easily and for children unable to take a tablet they disperse in a small amount of water.</p>