GUIDELINE

Cystic Fibrosis - Paediatric

Scope (Staff):	Clinical Staff – Medical, Nursing, Pharmacy
Scope (Area):	Perth Children's Hospital (PCH)

Child Safe Organisation Statement of Commitment

CAHS commits to being a child safe organisation by applying the National Principles for Child Safe Organisations. This is a commitment to a strong culture supported by robust policies and procedures to reduce the likelihood of harm to children and young people.

This document should be read in conjunction with this disclaimer

- These are empiric guidelines. Treatment may require modification due to known microbiology, previous treatment response or allergy status. Please contact infectious diseases to discuss treatment at any stage.
- In patients who receive aminoglycoside (IV or inhaled) treatment, audiology should be conducted as an outpatient after the admission to monitor for adverse effects. Additional assessment is required if there is onset of tinnitus or an increase in the severity of tinnitus.
- All patients should receive the annual influenza vaccine

CLINICAL SCENARIO	Usual duration	DRUGS/DOSES Standard Protocol	Monitoring
Exacerbation of cystic fibrosis Children ≥ 4 weeks of age	10 – 14 days	IV ceftazidime 50 mg/kg/dose (to a maximum of 3 grams) 8 hourly **OR** IV piperacillin/tazobactam 100 mg/kg/dose (to a maximum of 4 grams piperacillin component) 8 hourly **OR** IV cefepime 50 mg/kg/dose (to a maximum of 2 grams) 8 hourly **STRONGLY CONSIDER ADDITION of** IV tobramycin 10 mg/kg/dose (to a maximum initial dose of 660 mg) once daily with subsequent doses based on area under the curve therapeutic drug monitoring up to a maximum of 15 mg/kg/dose or 750 mg once daily (whichever is less). Ensure patients are well hydrated prior to commencement Refer to individual ChAMP monographs for Hospital in the Home (HITH) suitability.	Tobramycin Area under the Curve (AUC) measurement with the **FIRST** or **SECOND** dose, after any dose change and weekly for duration of treatment. Weekly Full Blood Count (FBC), Electrolytes, Urea and Creatinine (EUC) and Liver Function Tests (LFTs). Monitoring for vestibular toxicity should occur whilst on IV tobramycin and audiology assessment following admission.

<table>
<tr><th rowspan="2">CLINICAL SCENARIO</th><th rowspan="2">Usual duration</th><th>DRUGS/DOSES</th><th rowspan="2">Monitoring</th></tr>
<tr><th>Standard Protocol</th></tr>
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<td>Eradication of Pseudomonas aeruginosa in a child with cystic fibrosis</td>
<td>Variable</td>
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Eradication should be considered for all patients on the first isolation of Pseudomonas aeruginosa or after previous negative cultures from a sputum sample or bronchoalveolar lavage (BAL).

As for treatment of exacerbation of cystic fibrosis (see indication above). This may be followed by a course of inhaled tobramycin with or without the addition of oral ciprofloxacin (see dosing below)

Inhaled tobramycin
Children 6 months to 2 years old 150 mg twice daily via nebuliser
Children 2 years and older 300 mg twice daily via nebuliser. OR
Children 6 years and older 112 mg (4 x 28 mg capsules) administered twice daily via dry powder inhaler
CONSIDER ADDING
Children ≥ 4 weeks of age: oral ciprofloxacin 15-20 mg/kg/dose (to a maximum of 750 mg) 12 hourly. Consider the following suggested dose bands below to allow rounding to the most appropriate tablet size.
<table>
<tr><th>Weight</th><th>Dose</th></tr>
<tr><td>< 6 kg</td><td>20 mg/kg twice daily (consider liquid formulation)</td></tr>
<tr><td>6 kg to < 8.4 kg</td><td>125 mg twice daily</td></tr>
<tr><td>8.4 kg to <11 kg</td><td>187.5 mg twice daily</td></tr>
<tr><td>11 kg to <14 kg</td><td>250 mg twice daily</td></tr>
<tr><td>14 kg to <17 kg</td><td>312.5 mg twice daily</td></tr>
<tr><td>17 kg to <22 kg</td><td>375 mg twice daily</td></tr>
<tr><td>22 kg to <28 kg</td><td>500 mg twice daily</td></tr>
<tr><td>28 kg to <35 kg</td><td>625 mg twice daily</td></tr>
<tr><td>≥ 35 kg</td><td>750 mg twice daily</td></tr>
</table>
Alternative regimens are available on discussion with Infectious Diseases and Respiratory for individual patients or refractory cases.
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<td>For children on courses of oral antibiotics beyond 2 weeks of therapy recommend FBC, EUC and LFTs be done at 2 weeks and repeated if abnormal.

ciprofloxacin: patients should be instructed to alert team if any symptoms of arthropathy (e.g. bone or joint symptoms)</td>
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CLINICAL SCENARIO	Usual duration	DRUGS/DOSES Standard Protocol	Monitoring
Eradication of Methicillin sensitive *Staphylococcus aureus* (MSSA) in a child with cystic fibrosis	10-14 days	Strongly consider attempting eradication of MSSA in patients if abundant growth of MSSA or in cases where the patients lung health is not tracking optimally. Children ≥ 4 weeks of age: oral cefalexin 20 mg/kg/dose (to a maximum of 750 mg) given every 8 hours **IN CONJUNCTION WITH** Decolonisation for patients and family as part of the eradication. Apply nasal mupirocin 2% ointment to nares twice daily for 5 days **AND** chlorhexidine 2% hand and body wash (for children ≥ 3 months old) as a daily body wash for **5 days**. Refer to: MRSA and MSSA Guidelines for Staphylococcal decolonisation for further information.	

CLINICAL SCENARIO	Usual duration	DRUGS/DOSES Standard Protocol	Monitoring
Eradication of Methicillin Resistant Staphylococcus aureus (MRSA) in a child with cystic fibrosis Children ≥ 4 weeks of age	10-14 days	All patients with cystic fibrosis who have MRSA isolated from sputum samples should attempt MRSA eradication as below. Oral cotrimoxazole 4 mg/kg/dose (to a maximum of 160 mg trimethoprim component) 12 hourly **IN CONJUNCTION WITH** Decolonisation for patients and family is recommended as part of the eradication. Apply nasal mupirocin 2% ointment to nares twice daily for 5 days **AND** chlorhexidine 2% hand and body wash (for children ≥ 3 months old) as a daily body wash for **5 days**. Refer to: MRSA and MSSA Guidelines for Staphylococcal decolonisation for further information. **AND CONSIDER adding** Oral rifampicin[a] 15 mg/kg/dose (to a maximum of 600 mg) once daily	FBC, EUC, LFTs only after two weeks on treatment (i.e. if course is repeated). Cease treatment if thrombocytopenia or hepatotoxicity occurs.
Prevention of frequent exacerbations or primary prophylaxis in a child with cystic fibrosis	Up to 12 months	**CONSIDER** Oral azithromycin as an anti-inflammatory agent: Children ≥ 4 weeks – 6 years: 10 mg/kg/dose (to a maximum of 250 mg) three times a week Children ≥ 6 years AND 25 – < 40 kg: 250 mg three times a week Children ≥ 6 years AND ≥ 40 kg: 500 mg three times a week **OR** Children ≥ 1 year: 30 mg/kg/dose (to a maximum of 1.5 grams) once a week Exclude non-tuberculosis mycobacterial infection prior to initiation	LFTs and FBC monitored every 3 months on extended courses.

CLINICAL SCENARIO	Usual duration	DRUGS/DOSES Standard Protocol	Monitoring
Allergic Broncho-pulmonary Aspergillosis (ABPA) Children ≥ 4 weeks of age		Diagnosis is based on both clinical findings and immunological investigations: • Aspergillus infection. Aspergillus isolated from a respiratory specimen or precipitins/IgG to *A. fumigatus* • Clinical signs/symptoms: clinical deterioration not attributable to another aetiology OR new abnormalities on chest imaging unresponsive to antibiotic therapy • Allergic sensitisation: Serum total IgE >500 international units/mL and positive skin prick testing (SPT) *OR* IgE to *A. fumigatus* **Once ABPA confirmed:** Systemic glucocorticoids as first line therapy. **For refractory or severe cases consider addition of:** Oral itraconazole[b] with therapeutic drug monitoring (adjunctive therapy or as a steroid sparing agent) in discussion with Infectious Diseases: Itraconazole[b] **(Lozanoc®)** capsules: 1.25 – 2 mg/kg/dose (to a maximum initial dose of 100 mg) twice daily. **OR** Itraconazole[b] **(SAS approval required)** liquid: 5 mg/kg/dose (to a maximum initial dose of 100 mg) twice daily **Note:** Itraconazole interacts with many medications including ivacaftor / lumacaftor. Contact Pharmacy for advice.	FBC, EUC and LFTs performed monthly. Itraconazole levels should be measured 7 days after commencing, after any dose change or after any change in the formulation. Target levels 1 – 4 mg/L. **Once the dose is stable, levels should be checked monthly.**

a. Rifampicin is a red/protected ChAMP agent. However, when used in accordance with this guideline, prescribers do not need approval from infectious diseases prior to prescribing.
b. The available itraconazole formulations (Lozanoc® capsules and SAS formualtions) are NOT interchangeable. Only Lozanoc® capsules are kept at PCH. ALL prescriptions should state the formulation and brand required. Lozanoc® is the preferred brand of capsule due to less variability of bioavailability.

Related CAHS internal policies, procedures and guidelines
Antimicrobial Stewardship Policy (PCH Website)
ChAMP Empiric Guidelines

References and related external legislation, policies, and guidelines

1. Chen Y, Charbonnier JP, Andrinopoulou ER, Sly PD, Stick SM, Tiddens H. Azithromycin reduces bronchial wall thickening in infants with cystic fibrosis. J Cyst Fibros. 2024;23(5):870–3.
2. Stick SM, Foti A, Ware RS, Tiddens H, Clements BS, Armstrong DS, et al. The effect of azithromycin on structural lung disease in infants with cystic fibrosis (COMBAT CF): a phase 3, randomised, double-blind, placebo-controlled clinical trial. Lancet Respir Med. 2022;10(8):776–84.
3. Valery PC, Morris PS, Byrnes CA, Grimwood K, Torzillo PJ, Bauert PA, et al. Long-term azithromycin for Indigenous children with non-cystic-fibrosis bronchiectasis or chronic suppurative lung disease (Bronchiectasis Intervention Study): a multicentre, double-blind, randomised controlled trial. The Lancet Respiratory Medicine. 2013;1(8):610-20.
4. Wilms E, Touw D, Heijerman HM, van der Ent C. Azithromycin maintenance therapy in patients with cystic fibrosis: A dose advice based on a review of pharmacokinetics, efficacy, and side effects. Pediatric Pulmonology. 2012;47(7):658-65.
5. Royal Australian College of General Practitioners, Pharmaceutical Society of Australia, Australasian Society of Clinical and Experimental Pharmacologists and Toxicologists. AMH: Children's Dosing Companion. Adelaide: Australian Medicines Handbook Pty Ltd; 2025.
6. Langton Hewer SC, Smith S, Rowbotham NJ, Yule A, Smyth AR. Antibiotic strategies for eradicating Pseudomonas aeruginosa in people with cystic fibrosis. Cochrane Database Syst Rev. 2023;6(6):Cd004197.
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8. Garinis AC, Poling GL, Rubenstein RC, Konrad-Martin D, Hullar TE, Baguley DM, et al. Clinical Considerations for Routine Auditory and Vestibular Monitoring in Patients With Cystic Fibrosis. Am J Audiol. 2021;30(3s):800–9.

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