Local Guideline

John Hunter Children's Hospital
CHILDREN, YOUNG PEOPLE AND FAMILIES

NSW GOVERNMENT Health
Hunter New England Local Health District

Inhalation Therapy with Dornase alfa (Pulmozyme) in JHCH

Sites where Local Guideline applies	All clinical areas in JHCH
This Local Guideline applies to:	
1. Adults	No
2. Children up to 16 years	Yes
3. Neonates – less than 29 days	Yes
Target audience	Clinical staff who provide care to cystic fibrosis patients.
Description	This document outlines the criteria for use of dornase alfa (Pulmozyme) and the safe administration of this medication.
National Standard	1,4

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Keywords	Children, cough, cystic fibrosis, dornase alfa, mucus, Pulmozyme, sputum.
Document registration number	
Replaces existing document?	Yes
Registration number and dates of superseded documents	13.40 Inhalation Therapy with Pulmozyme® (dornase alfa) in JHCH

Related Legislation, Australian Standard, NSW Ministry of Health Policy Directive or Guideline, National Safety and Quality Health Service Standard (NSQHSS) and/or other, HNE Health Document, Professional Guideline, Code of Practice or Ethics:

- NSW Ministry of Health Policy Directive PD 2017_013 Infection Prevention and Control Policy

Local Guideline note	This document reflects what is currently regarded as safe and appropriate practice. The guideline section does not replace the need for the application of clinical judgment in respect to each individual patient but the procedure/s **require mandatory compliance**. If staff believe that the procedure/s should not apply in a particular clinical situation they must seek advice from their unit manager/delegate and document the variance in the patient's health record.
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This document contains advice on therapeutics	Yes Approval gained from Local Quality Use of Medicines Committee on 15/05/2018.
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Note: Over time, links in this document may cease working. Where this occurs, please source the document in the PPG Directory at: http://ppg.hne.health.nsw.gov.au/

PURPOSE AND RISKS

This procedure outlines the process for initiating treatment with dornase alfa (Pulmozyme) in the John Hunter Children's Hospital Paediatric Cystic Fibrosis clinic. Risks include incorrect prescription of medication and incorrect equipment provision.
Risk Category*:* Clinical Care & Patient Safety

GLOSSARY

Acronym or Term	Definition
PBS	Pharmaceutical Benefit Schedule

GUIDELINE

This Guideline does not replace the need for the application of clinical judgment in respect to each individual patient.

Staff Preparation

It is mandatory for staff to follow relevant: "Five moments of hand hygiene", infection control, moving safely/safe manual handling, documentation practices and to use HAIDET for patient/carer communication: **H**and hygiene **A**cknowledge, **I**ntroduce, **D**uration, **E**xplanation, **T**hank you or closing comment.

RATIONALE FOR TREATMENT

Dornase alfa (Pulmozyme) is an enzyme that breaks down the DNA in mucus, making it thinner and easier to cough up. Dornase alfa can improve lung function, reduce risk of lung infection, decrease need for hospital admissions and improve quality of life for patients with cystic fibrosis.

PATIENT SELECTION CRITERIA (PBS REQUIREMENT)

Treatment of cystic fibrosis in a patient who satisfies all the criteria as per PBS listing- refer to the PBS website and review criteria by clicking on "Authority required STREAMLINED"

TRIAL FOR PATIENTS OVER 5 YEARS

1. Baseline pulmonary function test during a stable period of the disease
2. 3-month trial of nebulised dornase alfa, at a dosage of 2.5 mg daily

Following 3-month trial: Review by respiratory doctor and independent CF team member and repeat pulmonary function test and global assessment. To be eligible for continued PBS-subsidised treatment:

1. The patient must demonstrate no deterioration in FEV_1 compared to baseline; AND
2. The patient or the patient's family must report improvement in airway clearance: AND
3. The treating specialist must report a benefit in the clinical status of the patient.

Patients should continue to have reviews every six months to establish that dornase alfa is continuing to produce worthwhile benefits.

TRIAL FOR PATIENTS UNDER 5 YEARS

Following an initial 6 months of therapy, a comprehensive assessment in CF clinic must be undertaken involving the patient, family, treating specialist and independent member of the CF team to establish agreement that dornase alfa is continuing to provide worthwhile benefit.

ADMINISTRATION

Dose

- 2.5 mg once a day (one nebule)

Delivery

- Via a PARI LC PLUS®/LC® Star nebuliser using a jet pump with an output 10 L/min or air from wall outlet > 6 L/min or personal pump e.g. eFlow®*rapid*, Aeroneb® Go

Long-term use

- It is recommended patients use dornase alfa on a daily basis. Lung function has been shown to improve during dornase alfa therapy but rapidly decline again when therapy is interrupted.
- Dornase alfa should be administered after airway clearance or at least half an hour before airway clearance. This should be individualised taking into consideration other inhaled medications and home routine.
- The PARI LC® Sprint nebuliser should be replaced every 6 months from supplies provided by Roche to the CF clinic.

Precautions: Patients should be reviewed by the cystic fibrosis team if acute haemoptysis occurs. If frank haemoptysis is significant, then dornase alfa should be suspended until reviewed by the cystic fibrosis medical team. Dornase alfa can be recommenced after 48 hours of no haemoptysis. Patients in remote or rural areas who experience frank haemoptysis should present to their local Emergency Department for assessment and management.

PROCEDURE

1. Pre-trial lung function test (for patients over 5 years)
2. Prescribe dornase alfa for appropriate time frame depending on age.

Age	Initiation Criteria	Continuation Criteria
> 5 yrs	FVC > 40% PLUS Evidence of chronic suppurative lung disease or obstruction **3-mth trial**	Post 3 mths initial therapy: No decline FEV_1 Improved airway clearance Clinical improvement
< 5 yrs	Severe clinical course > 3 admissions/y OR CF bronchiolitis with persistent wheeze OR Bronchiectasis on HRCT OR Physiological deficit reduced FOT or LCI **6-mth trial**	Post 6 mths: Clinical benefit

3. Educate patient & family in administration procedure & cleaning procedure.
4. Loan equipment—the CF clinic will loan a pump for the trial and will supply the nebuliser (PARI LC® Sprint)
5. Book patient for lung function test and doctor review three months following the commencement of treatment to assess response if over 5 years of age (patients under 5 years will be comprehensively assessed in CF clinic for level of clinical benefit).
6. Ongoing supply of the drug will continue through pharmacy, provided S100 eligibility criteria are met. Provide prescription for 1 month supply with 5 repeats. Ongoing supply of the equipment

- The PARI LC® Sprint nebuliser will be replaced every 6 months by the clinic. It is the patient's responsibility to clean and disinfect as per the manufacturer's guidelines.
- The pump is the patient's responsibility if purchased by the family, including maintenance, replacing filters and fault repair. If the pump is on loan from the clinic, servicing will be attended by biomedical engineering upon return of the pump every six months.

7. All patients should have a comprehensive assessment every 6 months to evaluate the benefit on ongoing treatment.

IMPLEMENTATION, MONITORING COMPLIANCE AND AUDIT

Guideline will be accessible to all staff in Cystic Fibrosis Clinic and new staff orientated to correct process.

REFERENCES

PBS dornase alfa