

Guideline and Procedure



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



Health
Hunter New England
Local Health District

Document Registration Number: 13.40

Inhalation Therapy with Dornase alfa (Pulmozyme®) in JHCH

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| Sites where Guideline and Procedure applies | All clinical areas in JHCH |
| Target audience: | Clinical staff who provide care to cystic fibrosis patients. |
| Description | |
| This Guideline and Procedure applies to: | |
| 1. Adults | No |
| 2. Children up to 16 years | Yes |
| 3. Neonates – less than 29 days | Yes |
| Keywords | Pulmozyme®, dornase alfa, cystic fibrosis, children, cough, mucus, sputum |
| Replaces Existing Guideline and Procedure | Yes |
| Registration Number(s) and/or name and of Superseded Documents | 13.40 Inhalation Therapy with Pulmozyme® (dornase alfa) in JHCH |
| Related Legislation, Australian Standards, NSW Ministry of Health Policy or Guideline, NSQHS Standard/EQuIP Criterion and/or other, HNE Health Documents, Professional Guidelines, Codes of Practice or Ethics: | <ul style="list-style-type: none"> NSW Ministry of Health Policy Directive 2007_079 Correct patient, Correct procedure, correct site http://www.health.nsw.gov.au/policies/pd/2007/pdf/PD2007_079.pdf NSW Ministry of Health Policy PD 2005_406 Consent to Medical Treatment http://www.health.nsw.gov.au/policies/PD/2005/pdf/PD2005_406.pdf NSW Ministry of Health Policy Directive PD 2007_036 Infection Control Policy http://www.health.nsw.gov.au/policies/pd/2007/pdf/PD2007_036.pdf |
| Guideline and Procedure 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 patients health record. |
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| This Guideline and Procedure contains advice on therapeutics | Yes Approval gained from HNE Quality Use of Medicines Committee on 13 th November 2014. |
| Date of Issue | 9 th December 2014 |
| Review due date | 9 th December 2017 |

RISK STATEMENT

This local clinical guideline and procedure has been developed to provide instruction to the healthcare worker and to ensure that the risks of harm to the patient associated with the use of Dornase alfa (Pulmozyme®) are identified and managed.

Any unplanned event resulting in, or with the potential for, injury, damage or other loss to the patient as a result of this clinical procedure must be reported through the Incident Information Management System. This would include unintended patient injury or complication from treatment that results in disability, death or prolonged hospital stay and is caused by health care management.

Open Disclosure procedures must be commenced to ensure the concerns of the patient are identified and managed in accordance with Ministry of Health Policy Directives. The Policy Directives and Guidelines for managing complaints and concerns about clinicians should be used in conjunction with other relevant NSW Health Policy Directives that govern the behaviour and actions of all staff.

RISK CATEGORY: Clinical Care & Patient Safety

OUTCOMES

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|---|--|
| 1 | This document outlines the criteria for use of Dornase alfa (Pulmozyme®) and the safe administration of this medication. |
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GUIDELINE

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

Summary

This procedure outlines the process for initiating treatment with dornase alfa (Pulmozyme®) in the John Hunter Children's Paediatric Cystic Fibrosis clinic.

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 www.pbs.gov.au/medicine/item/5704F-6120D and review criteria by clicking on "Authority required STREAMLINED"

Patients must be assessed at a cystic fibrosis clinic which is under the control of specialist respiratory specialists.

In patients who are 5 years of age or older:

- (1) have a FVC greater than 40% predicted for age, gender and height;
- (2) have evidence of chronic suppurative lung disease (cough and sputum most days of the week, or greater than 3 respiratory tract infections of more than 2 weeks' duration in any 12 months, or objective evidence of obstructive airways disease);

OR

In a patient less than 5 years of age who has:

- (1) A severe clinical course with frequent respiratory exacerbations or chronic respiratory symptoms (including chronic or recurrent cough, wheeze or tachypnoea) requiring hospital admissions more frequently than 3 times per year; OR
- (2) Significant bronchiectasis on chest high resolution computed tomography scan; OR
- (3) Severe cystic fibrosis bronchiolitis with persistent wheeze non-responsive to conventional medicines; or
- (4) Severe physiological deficit measure by forced oscillation technique or multiple breath nitrogen washout and failure to respond to conventional therapy.

Trial for patients over 5 years:

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

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 FEV1 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 alpha is continuing to produce worthwhile benefits.

Trial for patients under 5 years:

Following an initial 6 months 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

- 2.5mg once a day (one nebule)

Delivery

- Via a Pari LC Plus/Star™ nebulizer using a jet pump with an output 10L/m or air from wall outlet >6L/m or personal pump e.g. E flow 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 individualized taking into consideration other inhaled medications and home routine.
- The Pari™ nebulizer 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.

| Authority Number | Age | Initiation Criteria | Continuation Criteria |
|------------------|-------|---|--|
| 4288 | >5yrs | FVC> 40% PLUS Evidence of chronic suppurative lung disease or obstruction 3 mth trial | Post 3 mths initial therapy: No decline FEV1 Improved airway clearance Clinical improvement |

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|------|--------|--|---|
| 4300 | < 5yrs | Severe clinical course- > 3 admissions/yr 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 |
| 4296 | >5yrs | | Started Dornase alfa < 5yrs age Evidence of ongoing benefit |
| 4298 | < 5yrs | | Must have started Dornase alfa prior to 2009 Evidence of ongoing 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
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. The patient will be charged a dispensing fee for each supply.
7. Ongoing supply of the equipment
 - The Pari™ nebulizer 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.
8. All patients should have a comprehensive assessment every 6 months to evaluate the benefit on ongoing treatment.

References:

<http://www.pbs.gov.au/medicine/item/5704F-6120D>

Dentice and Elkins (2011) Timing of dornase alfa inhalation for cystic fibrosis. Cochrane Library

Consultation:

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APPROVED

CPGAG: June 2014

JHH QUM: November 2014

JHCH CQ&PCC: December 2014