

**Advice for patients receiving Enzyme Replacement Therapies (ERT) outside of Hospital
May 2020**

UK & Ireland LSD Nurse Collaboration

Clinical Centre :
Contact number:
Available hours :

Homecare team:
Contact number:
Available hours :

If the person feels unwell prior to having ERT, don't start infusion and seek advice from Clinical Team during working hours

If the person having ERT feels unwell during an ERT infusion or develops a rash

STOP THE INFUSION

Why does the person feel unwell? What is wrong?

Patients/carers who are semi-independent or fully independent with ERT infusions will be trained what to do in the event of an infusion-related reaction.

Patients should have a supply of supportive medications at home in order to treat the symptoms of a mild reaction as directed by the clinical team.

Mild Reaction

Flushing Itch Fever/chills

Runny nose Headache

Feeling sick "Not feeling right"

Moderate Reaction

Cough Uncontrollable shaking/shivering

Wheeze Fast heartbeat Chest tightness

Stomach cramps Vomiting/diarrhoea

Sudden loss or change in colour (paleness)

Widespread red rash or hives Agitation

Headache

Severe reaction

Difficulty in breathing (tongue swelling, gasping, swollen throat)

Feeling of abnormal heart rate/ palpitations

Chest pain

Uncontrolled defecation or vomiting

Swelling of the face/neck

Feeling faint/"passing out"

Dizziness Severe headache

Uncontrollable Shaking/shivers

Telephone the clinical team for advice

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CALL 999/112 IMMEDIATELY

If the clinical team recommends restarting the infusion and the symptoms persist, STOP the infusion

Telephone the clinical team to discuss before the next scheduled infusion.

A homecare nurse may need to visit for the next scheduled infusion.

If the clinical team recommends restarting the infusion and the symptoms persist, STOP the infusion

Telephone the clinical team to discuss before the next scheduled infusion.

A homecare nurse may need to visit for the next scheduled infusion, or it may need to take place in hospital.

A relative or friend should stay with the person until they are transferred by ambulance to hospital.

The paramedics will give medication to help with the reaction.

The local hospital must contact your clinical team for advice.

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Your Clinical Team will advise which of these medicines should be kept at home.

Advice may be to:

- Take oral paracetamol to treat symptoms of fever, chills or headache
- Take an oral antihistamine for symptoms of flushing, itching or runny nose

Please note, if any of these medicines are administered within 4 hours of starting the infusion, **do not repeat the dose** before discussing with a healthcare professional

Paracetamol

From age 6 months up to 6 years: Paracetamol 120mg/5mL oral solution

Age	Volume	Number of times per day
6-23 months	5mL	Every 4-6 hours Maximum 4 times per day
2-3 years	7.5mL	
4-5 years	10mL	

From age 6 years up to 12 years: Paracetamol 250mg/5mL oral solution

Age	Volume	Number of times per day
6-7 years	5mL	Every 4-6 hours Maximum four times per day
8-9 years	7.5mL	
10-11 years	10mL	

Age 12+ years: Paracetamol 500mg tablets

Age	Number of tablets	Number of times per day
12-15 years	1 - 1.5 tablets	Every 4-6 hours Maximum four times per day
16 years +	2 tablets	

Loratadine (non-sedating antihistamine)

From age 2 years up to 12 years: Loratadine 5mg/mL oral solution

Age	Volume	Number of times per day
2-11 years (up to 31kg)	5mL	Once daily
2-11 years (31kg and above)	10mL	

Age 12+ years: Loratadine 10mg tablets

Age	Number of tablets	Number of times per day
12 years +	1 tablet	Once daily

Ibuprofen

From age 6 months up to 12 years: Ibuprofen 100mg/5mL oral solution

Age	Volume	Number of times per day
6-11 months	2.5mL	Every 6-8 hours Maximum three times per day
1-3 years	5mL	
4-6 years	7.5mL	
7-9 years	10mL	
10-11 years	15mL	

Age 12+ years: Ibuprofen 400mg tablets

Age	Number of tablets	Number of times per day
12 years +	1 tablet	Every 6-8 hours Maximum three times per day

Chlorphenamine (sedating antihistamine)

From age 6 months up to 12 years: Chlorphenamine 2mg/5mL oral solution

Age	Volume	Number of times per day
6-23 months	2.5mL	Maximum two times per day
2-5 years	2.5mL	Every 4-6 hours Maximum six times per day
6-11 years	5mL	Every 4-6 hours Maximum six times per day

Age 12+ years: Chlorphenamine 4mg tablets

Age	Number of tablets	Number of times per day
12 years +	1 tablet	Every 4-6 hours Maximum six times per day

Cetirizine (non-sedating antihistamine)

From age 2 years up to 12 years: Cetirizine 1mg/mL oral solution

Age	Volume	Number of times per day
2-5 years	2.5mL	Maximum two times per day
6-11 years	5mL	

Age 12+ years: Cetirizine 10mg tablets

Age	Number of tablets	Number of times per day
12 years +	1 tablet	Once daily