Fax this completed form to: 1-888-990-0008 Phone: 1-866-888-0660

ELAPRASE® START Form and Authorization for Shire OnePath® Services

Please see the Indication and Important Safety Information, including Boxed Warning regarding Risk of Anaphylaxis, below and Full Prescribing Information.

elaprase® (idursulfase) OnePath®

1 Prescribing Physician Information

Name (First, Last)

Street Address | City

State | Zip Code | Telephone ( ) | Fax ( )

Office Contact | Tax ID #

State Licence # | National Provider ID #

2 Site of Care Information

Site of Care Name | ☐ Home Infusion (provide address of Home Infusion Company below)

Street Address | City

State | Zip Code | Telephone ( ) | Fax ( )

Office Contact | National Provider ID #

3 Patient Information

First Name | Middle Initial | ☐ Male ☐ Female

Last Name | DOB: Month/Day/Year

Age | Last 4 digits of SSN | Patient weight (kg)

Street Address | City

State | Zip Code | Mobile Telephone ( ) | Email Address

Home Telephone ( ) | Work Telephone ( ) | Caregiver Telephone ( )

Caregiver Name (First, Last) | Relationship to Patient

4 Insurance Information

Please attach copies of both sides of patient's insurance card(s)

☐ Check if patient does not have insurance

Primary Insurance | Insurance Telephone ( ) | Policy ID # | Group #

Policy Holder Name (First, Last) | Relationship to Patient

Pharmacy Plan Name | Rx Bin #

Pharmacy Plan Telephone ( ) | Rx PCN #

Secondary Insurance | Insurance Telephone ( ) | Policy ID # | Group #

Policy Holder Name (First, Last) | Relationship to Patient

5 Physician Authorization to Initiate ELAPRASE®

☐ ELAPRASE® (Box must be checked to initiate therapy)

I appoint Shire, its affiliates and their representatives (collectively "Shire") to convey on my behalf the prescription described herein to a pharmacy, if applicable.

Sign here

Prescriber Signature: _____

(STAMPS NOT ACCEPTABLE) DISPENSE AS WRITTEN

Date: _____

6 Patient Authorization to Share Personal Health Information and OnePath® Enrollment

☑ I authorize any health plan, physician, health care professional, hospital, clinic, pharmacy provider or other health care provider (collectively, "Health Care Providers") to disclose my personal health information, including information relating to my medical condition, treatment, care management, and health insurance, as well as all information provided on this form and any prescription, personal health information obtained by Health Care Providers prior to the date of this authorization ("Personal Health Information"), to Shire Human Genetic Therapies, Inc., its affiliates and their representatives, agents, and contractors (collectively, "Shire") and to receive financial remuneration from Shire in exchange, for the following purposes: for Shire to provide product support services, including coordination of benefits and therapy; reimbursement support; investigating insurance coverage; communicating with me by mail, email, or telephone about my medical condition, treatment, care management, and health insurance; and internal use by Shire, including data analysis. I understand that my Personal Health Information disclosed under this authorization may be redisclosed by Shire and no longer protected by federal privacy laws. I understand, however, that Shire agrees to undertake reasonable efforts to maintain my Personal Health Information in a secure manner and not to disclose it to third parties without a legitimate reason for doing so.
I understand that I may refuse to sign this Authorization and that my treatment, payment, enrollment or eligibility for benefits, including my access to therapy, is not conditioned on my signing this Authorization. I understand that I am entitled to a signed copy of this Authorization. This Authorization expires one year from the date of execution, or one year after the date of my last prescription, whichever is later. I understand that I may revoke this Authorization at any time by sending written notice of revocation to OnePath®, 300 Shire Way, Lexington, MA 02421, which becomes effective upon receipt by any Health Care Provider subject to federal privacy laws, except to the extent that action already has been taken in reliance on this Authorization.

Check here

☐ ***OnePath® Enrollment*** (must check box to be enrolled in product support services through OnePath®)
I certify that all of the information provided on this form is complete and accurate. I authorize Shire to collect Personal Health Information from me, my caregivers, and Health Care Providers, and to use and disclose such Personal Health Information to provide product support services, including but not limited to coordination of benefits and therapy; reimbursement support; investigating insurance coverage; communicating with me by mail, email, or telephone about my medical condition, treatment, care management, and health insurance.

Sign here

Patient Signature: _____

Date: _____

Legal Representative Signature (if applicable): _____

Date: _____

INDICATIONS AND USAGE

ELAPRASE® (Idursulfase) is indicated for patients with Hunter syndrome (Mucopolysaccharidosis II, MPS II). ELAPRASE has been shown to improve walking capacity in patients 5 years and older.

In patients 16 months to 5 years of age, no data are available to demonstrate improvement in disease-related symptoms or long term clinical outcome; however, treatment with ELAPRASE has reduced spleen volume similarly to that of adults and children 5 years of age and older.

The safety and efficacy of ELAPRASE have not been established in pediatric patients less than 16 months of age.

IMPORTANT SAFETY INFORMATION

WARNING: RISK OF ANAPHYLAXIS

Life-threatening anaphylactic reactions have occurred in some patients during and up to 24 hours after ELAPRASE infusions. Anaphylaxis, presenting as respiratory distress, hypoxia, hypotension, urticaria and/or angioedema of throat or tongue have been reported to occur during and after ELAPRASE infusions, regardless of duration of the course of treatment. Closely observe patients during and after ELAPRASE administration and be prepared to manage anaphylaxis. Inform patients of the signs and symptoms of anaphylaxis and have them seek immediate medical care should symptoms occur. Patients with compromised respiratory function or acute respiratory disease may be at risk of serious acute exacerbation of their respiratory compromise due to hypersensitivity reactions, and require additional monitoring.

- **Hypersensitivity Reactions Including Anaphylaxis:** Ensure that personnel administering product are adequately trained in cardiopulmonary resuscitative measures, and have ready access to emergency medical services (EMS).

  If anaphylactic or other acute reactions occur, immediately discontinue the infusion of ELAPRASE and initiate appropriate medical treatment. Observe patients closely for an appropriate period of time after administration of ELAPRASE, taking into account the time to onset of anaphylaxis seen in premarketing clinical trials and postmarketing reports. Inform patients of the signs and symptoms of anaphylaxis, and instruct them to seek immediate medical care should signs and symptoms occur. When severe reactions have occurred during clinical trials, subsequent infusions were managed with antihistamine and/or corticosteroids prior to or during infusions, a slower rate of ELAPRASE infusion, and/or early discontinuation of the ELAPRASE infusion.

- **Risk of Hypersensitivity, Serious Adverse Reactions, and Antibody Development in Hunter Syndrome Patients with Severe Genetic Mutations:** Hunter syndrome patients aged 7 years and younger with complete gene deletion, large gene rearrangement, nonsense, frameshift or splice site mutations experienced a higher incidence of hypersensitivity reactions, serious adverse reactions and anti-idursulfase antibody development.

- **Risk of Acute Respiratory Complications:** Patients with compromised respiratory function or acute febrile or respiratory illness may be at higher risk of life-threatening complications from hypersensitivity reactions. Careful consideration should be given to the patient's clinical status prior to administration of ELAPRASE and consider delaying the ELAPRASE infusion.

- **Risk of Acute Cardiorespiratory Failure:** Caution should be exercised when administering ELAPRASE to patients susceptible to fluid overload, or patients with acute underlying respiratory illness or compromised cardiac and/or respiratory function for whom fluid restriction is indicated. These patients may be at risk of serious exacerbation of their cardiac or respiratory status during infusions. Appropriate medical support and monitoring measures should be readily available during ELAPRASE infusion, and some patients may require prolonged observation times that should be based on the individual needs of the patient.

- **Adverse Reactions:** In clinical trials, the most frequent serious adverse reactions following ELAPRASE treatment were hypoxic episodes. Other notable serious adverse reactions that occurred in the ELAPRASE treated patients but not in the placebo treated patients included one case each of: cardiac arrhythmia, pulmonary embolism, cyanosis, respiratory failure, infection, and arthralgia.

  The most common adverse reactions occurring in at least three patients (≥9%) aged five years and older were headache, pruritus, musculoskeletal pain, urticaria, diarrhea, and cough. The most common adverse reactions occurring in at least three patients (≥10%) aged seven years and younger were pyrexia, rash, vomiting, and urticaria. In all clinical trials, the most common adverse reactions requiring medical intervention were hypersensitivity reactions, and included rash, urticaria, pruritus, flushing, pyrexia, and headache.

- **Immunogenicity:** In clinical trials in patients 5 years and older, 32 of 63 (51%) patients tested positive for anti-idursulfase IgG antibodies (Ab) at least one time. Of the 32 Ab-positive patients, 23 of 32 (72%) tested positive for Ab at three or more different time points (persistent Ab). The incidence of hypersensitivity reactions was higher in patients who tested positive for Ab than those who tested negative.

  Thirteen of 32 (41%) Ab-positive patients also tested positive for antibodies that neutralize idursulfase uptake into cells (neutralizing antibodies, NAb) or enzymatic activity at least one time, and 8 (25%) of Ab-positive patients had persistent NAb. There was no clear relationship between the presence of either Ab or NAb and therapeutic response.

  In the clinical trial in patients 7 years and younger, 19 of 28 (68%) patients treated with ELAPRASE 0.5 mg/kg once weekly tested Ab-positive, with 16 of 19 (84%) having persistent Ab. In addition, 15 of 19 (79%) Ab-positive patients tested positive for NAb, with 14 of 15 (93%) having persistent NAb.

- **Postmarketing Experience:** Late-emergent symptoms and signs of anaphylactic reactions have occurred up to 24 hours after initial treatment and recovery from an initial anaphylactic reaction. In addition, patients experienced repeated anaphylaxis over a two to four month period, up to several years after initiating ELAPRASE treatment.

  Serious adverse reactions that resulted in death included cardiorespiratory arrest, respiratory failure, respiratory distress, cardiac failure, and pneumonia.

Please click here for Full Prescribing Information, including Boxed Warning

To report SUSPECTED ADVERSE REACTIONS, contact Shire Medical Information at 1-866-888-0660 or FDA at 1-866-FDA-1088 or www.fda.gov/medwatch

Please fax this form to: 1-888-990-0008