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NOVA SCOTIA

PharmacareNEWS

inside

Nova Scotia Formulary Updates

New Exception Status Benefits

The following products have been listed with the following criteria, effective **immediately**.

PRODUCT	STRENGTH	DIN	PRESCRIBER	BENEFIT STATUS	MFR
Elelyso (taliglucerase alfa)	200U/Vial Pws for Inj	02425637	DNP	E (SF)	PFI
Criteria	• For the treatment of patients with symptomatic Gaucher disease type 1 (GD1) for whom treatment with velaglucerase alfa is not tolerated or contraindicated. **Clinical Notes:** • Velaglucerase alfa is the preferred reimbursed enzyme replacement therapy for GD1. • Requests for patients currently using taliglucerase alfa who do not have a contraindication or intolerance to velaglucerase alfa will be considered for coverage of velaglucerase alfa only. • Requests for coverage must meet the criteria for diagnosis of GD1, indication for therapy and expected response to enzyme replacement therapy outlined below: **Initial Coverage** **Diagnosis** • The diagnosis of GD1 must have been established by the demonstration of specific deficiency of glucocerebrosidase (GCase) in tissue or cultured skin fibroblasts, or by demonstration of the presence, in tissue or peripheral blood leukocytes, of mutations in the GCase gene known to result in severe enzyme deficiency.				

New Exception Status Benefits Continued…

PRODUCT	STRENGTH	DIN	PRESCRIBER	BENEFIT STATUS	MFR
Elelyso (taliglucerase alfa)	200U/Vial Pws for Inj	02425637	DNP	E (SF)	PFI
Criteria	• Other potentially confounding diagnoses, such as Hodgkin disease or other storage disorders, must have been ruled out. The symptoms experienced by the patient should be shown to be attributable to GD1 and not another condition that might mimic it. • The patient should not have any GD1-related or other medical condition that might reasonably be expected to compromise their response to treatment. In some patients with GD1, secondary pathologic changes, such as avascular necrosis of bone, may already have occurred that would not be expected to respond to enzyme replacement. In such patients, reversal of the pathology is unlikely. **Disease Severity** Evidence of disease severity must be provided, and include at least one of the following: • **Hematological complications** o Hemoglobin <85% of lower limit of age- and sex-appropriate normal after other causes of anemia, such as iron deficiency, have been treated or ruled out. o Platelet count <50 x 10^9/L on two separate occasions at least one month apart. Higher cut offs may be considered in the event the patient is symptomatic with bleeding or bruising. o At least two episodes of severely symptomatic splenic infarcts confirmed by CT or other imaging of the abdomen. • **Skeletal complications** o A single acute bone crisis severe enough to require hospitalization or marked incapacitation. o Radiographic or MRI evidence of incipient destruction of any major joint (e.g., hips and shoulders) or significant worsening of bony pathology (e.g. marrow infiltration, avascular necrosis, and infarcts). o Spontaneous fractures with evidence from imaging studies that recurrence is likely. o Chronic bone pain causing significant loss of time from work or school and not controlled by administration of non-narcotic analgesics or anti-inflammatory drugs. o Note: Patients who are scheduled for major joint replacement surgery, made necessary by skeletal complications of GD1, should be treated with enzyme therapy at a dosage of at least 30 units/kg every 2 weeks for at least 6 months before the joint replacement surgery and the dose continued until rehabilitation from the surgery is complete.				

New Exception Status Benefits Continued…

PRODUCT	STRENGTH	DIN	PRESCRIBER	BENEFIT STATUS	MFR
Elelyso (taliglucerase alfa)	200U/Vial Pws for Inj	02425637	DNP	E (SF)	PFI

Criteria	• **Gastrointestinal complications**
	  ○ Evidence of significant liver dysfunction attributable to GD1, such as portal hypertension or impaired hepatic synthetic function. Elevation of transaminase levels with no evidence of portal hypertension or impairment in synthetic function is not an indication for ERT.
	  ○ Significant discomfort due to enlargement of the spleen or liver.
	• **Pulmonary complications**
	  ○ Evidence of clinically significant and/or progressive pulmonary disease due to GD1.
	• **Systemic complications**
	  ○ Growth failure in children: significant decrease in percentile linear growth over a 3 - 6 month period.
	Exclusion Criteria
	• Due to the absence of data demonstrating therapy of asymptomatic patients alters long term outcomes, asymptomatic patients will not be considered for coverage.
	• Data does not suggest that ERT is effective in improving central nervous system involvement in patients with Type 2 and 3 disease. Therefore, patients exhibiting primary neurological disease due to GD1 will not be considered for coverage. Treatment for patients at risk of neuronopathic disease should be guided by the non-neurological manifestations of their disease as outlined above and ERT should not be initiated in asymptomatic patients who have a genotype that increases their risk of neuronopathic involvement.
	Continued Coverage
	• Patients' disease severity must be re-assessed annually.
	• A patient may receive approval for further coverage for treatment where there is demonstrated clinical improvement based on the expected response outlined below:				

New Exception Status Benefits Continued…

PRODUCT	STRENGTH	DIN	PRESCRIBER	BENEFIT STATUS	MFR
Elelyso (taliglucerase alfa)	200U/Vial Pws for Inj	02425637	DNP	E (SF)	PFI

Criteria	Indication for therapy	Expected Response
	Hemoglobin < 85% of lower limit of age and sex-appropriate normal	Increase hemoglobin levels to > 110 for women and children and > 120 for men
	Platelet count < 50 x 10^9/L on two separate occasions, or bleeding complications associated with thrombocytopenia irrespective of the platelet count	Increase platelet count to level sufficient to prevent spontaneous bleeding
		Normalization of platelet count in splenectomized patients
		In patients with intact spleen, an increase of at least 1.5X baseline value
	Two episodes of severely symptomatic splenic infarcts	Reduction of spleen volume by 50%
		Prevention of further splenic infarcts
	Acute bone crises	Prevent bone crises
	Radiographic or MRI evidence of incipient destruction of any major joint	Improvement in imaging parameters (either MRI, QCSI[1], or BMD)
	Spontaneous fractures	Prevention of further fractures
	Chronic bone pain	Reduce bone pain
	Major joint replacement surgery	Optimize surgical outcome
	Significant hepatic dysfunction	Improvement in hepatic function
	Symptomatic hepatosplenomegaly	Reduction of spleen volume by 50%
		Reduction in liver volume by 30%

New Exception Status Benefits Continued…

PRODUCT	STRENGTH	DIN	PRESCRIBER	BENEFIT STATUS	MFR
Elelyso (taliglucerase alfa)	200U/Vial Pws for Inj	02425637	DNP	E (SF)	PFI

Criteria	Indication for therapy	Expected Response
	Progressive pulmonary disease due to GD1	Improvement in pulmonary hypertension[2]
		Improvement in oxygenation
		Reversal of hepatopulmonary syndrome
	Growth failure in children	Return to normal range of growth parameters

1. QCSI- quantitative chemical shift imaging
2. May require adjuvant treatment for pulmonary hypertension

Discontinuation of Coverage

- Renewals will NOT be approved if:
  - The patient or the patient's specialist fails to comply adequately with treatment or measures taken to evaluate the effectiveness of the therapy (e.g. monitoring for expected response).
  - Therapy fails to relieve the symptoms of disease that originally resulted in the patient being approved for treatment.

Claim Notes:

- Approvals will be for a maximum of 60 units/kg every 2 weeks.
- Initial Approval: 6 months.
- Renewal Approval: 1 year.
- Claims that exceed the maximum claim amount of $9,999.99 must be divided and submitted as separate transactions using the DIN first and then the following PINs:
  - 00904383
  - 00904385

PRODUCT	STRENGTH	DIN	PRESCRIBER	BENEFIT STATUS	MFR
Mavenclad (cladribine)	10mg Tab	02470179	DNP	E (SF)	EMD

Criteria

- For the treatment of adult patients with relapsing-remitting multiple sclerosis (RRMS) who meet all the following criteria:
  - Confirmed diagnosis based on McDonald criteria.
  - Has experienced one or more disabling relapses or new MRI activity in the past year.

New Exception Status Benefits Continued…

PRODUCT	STRENGTH	DIN	PRESCRIBER	BENEFIT STATUS	MFR
Mavenclad (cladribine)	10mg Tab	02470179	DNP	E (SF)	EMD
Criteria	○ Ambulatory with or without aid (i.e. has a recent Expanded Disability Status Scale (EDSS) score of less than or equal to 6.5). ○ Refractory or intolerant to at least one disease modifying therapy (e.g., interferon, glatiramer, dimethyl fumarate, teriflunomide, ocrelizumab). **Clinical Notes:** • Treatment should be discontinued for patients with an EDSS score of greater than or equal to 7. • A relapse is defined as the appearance of new or worsening neurological symptoms in the absence of fever or infection, lasting at least 24 hours yet preceded by stability for at least one month and accompanied by new objective neurological findings observed through evaluation by a neurologist. **Claim Notes:** • Must be prescribed by a neurologist with experience in the treatment of multiple sclerosis. • Approvals will be for 1.75mg/kg to a maximum of 200mg per treatment year. • Approval period: 2 years. • Claims that exceed the maximum claim amount of $9,999.99 must be divided and submitted as separate transactions using the DIN first and then the following PINs: ○ 00904524 ○ 00904525 ○ 00904526				

PRODUCT	STRENGTH	DIN	PRESCRIBER	BENEFIT STATUS	MFR
Mictoryl (propiverine hydrochloride)	5mg Tab	02460289	DNP	E (F)	DUI
Criteria	• For the treatment of overactive bladder with symptoms of urgency incontinence and/or urinary frequency and urgency in pediatric patients under 18 years of age.				

New Exception Status Benefits Continued…

PRODUCT	STRENGTH	DIN	PRESCRIBER	BENEFIT STATUS	MFR
VPRIV (velaglucerase alfa)	400U/Vial Pws for Inj	02357119	DNP	E (SF)	SHI

Criteria

- For the treatment of patients with symptomatic Gaucher disease type 1 (GD1) for whom treatment with velaglucerase alfa is tolerated or not contraindicated.

Clinical Notes:

- Velaglucerase alfa is the preferred reimbursed enzyme replacement therapy (i.e. first tier) for all new and existing GD1.
- Requests for patients currently using taliglucerase alfa who do not have a contraindication or intolerance to velaglucerase alfa will be switched to velaglucerase alfa only.
- Requests for coverage must meet the criteria for diagnosis of GD1, indication for therapy and expected response to enzyme replacement therapy outlined below:

Initial Coverage

Diagnosis

- The diagnosis of GD1 must have been established by the demonstration of specific deficiency of glucocerebrosidase (GCase) in tissue or cultured skin fibroblasts, or by demonstration of the presence, in tissue or peripheral blood leukocytes, of mutations in the GCase gene known to result in severe enzyme deficiency.
- Other potentially confounding diagnoses, such as Hodgkin disease or other storage disorders, must have been ruled out. The symptoms experienced by the patient should be shown to be attributable to GD1 and not another condition that might mimic it.
- The patient should not have any GD1-related or other medical condition that might reasonably be expected to compromise their response to treatment. In some patients with GD1, secondary pathologic changes, such as avascular necrosis of bone, may already have occurred that would not be expected to respond to enzyme replacement. In such patients, reversal of the pathology is unlikely.

Disease Severity

Evidence of disease severity must be provided, and include at least one of the following:

- **Hematological complications**
  - Hemoglobin <85% of lower limit of age- and sex-appropriate normal after other causes of anemia, such as iron deficiency, have been treated or ruled out.
  - Platelet count <50 x 10^9/L on two separate occasions at least one month apart. Higher cut offs may be considered in the event the patient is symptomatic with bleeding or bruising.
  - At least two episodes of severely symptomatic splenic infarcts confirmed by CT or other imaging of the abdomen.
- **Skeletal complications**
  - A single acute bone crisis severe enough to require hospitalization or marked incapacitation.

New Exception Status Benefits Continued…

PRODUCT	STRENGTH	DIN	PRESCRIBER	BENEFIT STATUS	MFR
VPRIV (velaglucerase alfa)	400U/Vial Pws for Inj	02357119	DNP	E (SF)	SHI

Criteria

- Radiographic or MRI evidence of incipient destruction of any major joint (e.g., hips and shoulders) or significant worsening of bony pathology (e.g. marrow infiltration, avascular necrosis, and infarcts).
- Spontaneous fractures with evidence from imaging studies that recurrence is likely.
- Chronic bone pain causing significant loss of time from work or school and not controlled by administration of non-narcotic analgesics or anti-inflammatory drugs.
- Note: Patients who are scheduled for major joint replacement surgery, made necessary by skeletal complications of GD1, should be treated with enzyme therapy at a dosage of at least 30 units/kg every 2 weeks for at least 6 months before the joint replacement surgery and the dose continued until rehabilitation from the surgery is complete.

- **Gastrointestinal complications**
  - Evidence of significant liver dysfunction attributable to GD1, such as portal hypertension or impaired hepatic synthetic function. Elevation of transaminase levels with no evidence of portal hypertension or impairment in synthetic function is not an indication for ERT.
  - Significant discomfort due to enlargement of the spleen or liver.
- **Pulmonary complications**
  - Evidence of clinically significant and/or progressive pulmonary disease due to GD1.
- **Systemic complications**
  - Growth failure in children: significant decrease in percentile linear growth over a 3 - 6 month period.

Exclusion Criteria

- Due to the absence of data demonstrating therapy of asymptomatic patients alters long term outcomes, asymptomatic patients will not be considered for coverage.
- Data does not suggest that ERT is effective in improving central nervous system involvement in patients with Type 2 and 3 disease. Therefore, patients exhibiting primary neurological disease due to GD1 will not be considered for coverage. Treatment for patients at risk of neuronopathic disease should be guided by the non-neurological manifestations of their disease as outlined above and ERT should not be initiated in asymptomatic patients who have a genotype that increases their risk of neuronopathic involvement.

Continued Coverage

- Patients' disease severity must be re-assessed annually.
  - A patient may receive approval for further coverage for treatment where there is demonstrated clinical improvement based on the expected response outlined below:

New Exception Status Benefits Continued…

PRODUCT	STRENGTH	DIN	PRESCRIBER	BENEFIT STATUS	MFR
VPRIV (velaglucerase alfa)	400U/Vial Pws for Inj	02357119	DNP	E (SF)	SHI

Criteria	Indication for therapy	Expected Response
	Hemoglobin < 85% of lower limit of age and sex-appropriate normal	Increase hemoglobin levels to > 110 for women and children and > 120 for men
	Platelet count < 50 x 10^9/L on two separate occasions, or bleeding complications associated with thrombocytopenia irrespective of the platelet count	Increase platelet count to level sufficient to prevent spontaneous bleeding
		Normalization of platelet count in splenectomized patients
		In patients with intact spleen, an increase of at least 1.5X baseline value
	Two episodes of severely symptomatic splenic infarcts	Reduction of spleen volume by 50%
		Prevention of further splenic infarcts
	Acute bone crises	Prevent bone crises
	Radiographic or MRI evidence of incipient destruction of any major joint	Improvement in imaging parameters (either MRI, QCSI[1], or BMD)
	Spontaneous fractures	Prevention of further fractures
	Chronic bone pain	Reduce bone pain
	Major joint replacement surgery	Optimize surgical outcome
	Significant hepatic dysfunction	Improvement in hepatic function
	Symptomatic hepatosplenomegaly	Reduction of spleen volume by 50%
		Reduction in liver volume by 30%
	Progressive pulmonary disease due to GD1	Improvement in pulmonary hypertension[2]
		Improvement in oxygenation
		Reversal of hepatopulmonary syndrome
	Growth failure in children	Return to normal range of growth parameters

1. QCSI- quantitative chemical shift imaging
2. May require adjuvant treatment for pulmonary hypertension

Discontinuation of Coverage

- Renewals will NOT be approved if:
  - The patient or the patient's specialist fails to comply adequately with treatment or measures taken to evaluate the effectiveness of the therapy (e.g. monitoring for expected response).
  - Therapy fails to relieve the symptoms of disease that originally resulted in the patient being approved for treatment.

New Exception Status Benefits Continued…

PRODUCT	STRENGTH	DIN	PRESCRIBER	BENEFIT STATUS	MFR
VPRIV (velaglucerase alfa)	400U/Vial Pws for Inj	02357119	DNP	E (SF)	SHI
Criteria	**Claim Notes:** • Approvals will be for a maximum of 60 units/kg every 2 weeks. • Initial Approval: 6 months. • Renewal Approval: 1 year. • Claims that exceed the maximum claim amount of $9,999.99 must be divided and submitted as separate transactions using the DIN first and then the following PINs: o 00904378 o 00904379 o 00904380				

Criteria Updates

The following criteria has been updated effective **immediately:**

PRODUCT	STRENGTH	DIN	PRESCRIBER	BENEFIT STATUS	MFR
Lenvima (lenvatinib)	4mg Compliance Pack	02484056	DNP	E (SFC)	EIS
	8mg Compliance Pack	02468220	DNP	E (SFC)	EIS
	12mg Compliance Pack	02484129	DNP	E (SFC)	EIS
Criteria	• For the first-line treatment of adult patients with unresectable or metastatic hepatocellular carcinoma who meet all the following criteria: o Child-Pugh class status of A. o ECOG performance status of 0 or 1. o Less than 50% liver involvement and no invasion of the bile duct or main portal vein. o No brain metastases or prior liver transplantation. **Clinical Notes:** • Treatment should be continued until disease progression or unacceptable toxicity. • Patients who are unable to tolerate lenvatinib may be switched to sorafenib if there is no disease progression and provided all other funding criteria are met. • Patients with disease progression on lenvatinib are not eligible for reimbursement of sorafenib.				

Criteria Updates Continued…

PRODUCT	STRENGTH	DIN	PRESCRIBER	BENEFIT STATUS	MFR
Nexavar (Sorafenib)	200mg Tab	02284227	DNP	E (SFC)	BAY
Criteria	• As a single agent first line systemic therapy option in adult patients with a diagnosis of hepatocellular carcinoma (HCC) with Child-Pugh Class A liver dysfunction (mild hepatic impairment) with ECOG performance status 0-1; and who have either progression of disease, or who are not candidates for curative intent treatments (transplantation, hepatic resection), or other well established palliative interventions (ablation, transcatheter arterial chemo-embolization (TACE), internal radiation). **Clinical Note:** • Patients who are unable to tolerate sorafenib may be switched to lenvatinib if there is no disease progression and provided all other funding criteria are met. • Patients with disease progression on sorafenib are not eligible for reimbursement of lenvatinib.				

PRODUCT	STRENGTH	DIN	PRESCRIBER	BENEFIT STATUS	MFR
Stivarga (Regorafenib)	40mg Tab	02403390	DNP	E (SFC)	BAY

Criteria	• For the treatment of patients with unresectable hepatocellular carcinoma (HCC) who have experienced disease progression on sorafenib or lenvatinib and meet all of the following criteria:
	  ○ Child-Pugh class status of A.
	  ○ ECOG performance status of 0 or 1.
	Clinical Note:
	• Treatment should continue until disease progression or unacceptable toxicity.
	• Patients with disease progression on sorafenib must have tolerated a minimum dose of 400 mg per day for at least 20 of the last 28 days of treatment.				

New Products

Effective **immediately,** the following new products have been added to the Nova Scotia Formulary. The benefit status within the Pharmacare Programs is indicated.

PRODUCT	STRENGTH	DIN	PRESCRIBER	BENEFIT STATUS	MFR
Nucala	100mg/mL Autoinjector	02492989	DNP	E (SF)	GSK
Nucala	100mg/mL Pre-filled Syringe	02492997	DNP	E (SF)	GSK
Vyzulta	0.024% Oph Sol	02484218	DNP	SF	BSL

New Form

New request form for Ocrevus can be found at the following link:

https://novascotia.ca/dhw/pharmacare/exception-status-drugs.asp

Legend

PRESCRIBER CODES	BENEFIT STATUS	MANUFACTURER CODES
D - Physician / Dentist	S - Seniors' Pharmacare	BAY - Bayer Inc.
N - Nurse Practitioner	F - Community Services Pharmacare	BSL - Bausch Health, Canada Inc.
P - Pharmacist	- Family Pharmacare	DUI - Duchesnay Inc.
M - Midwife	C - Drug Assistance for Cancer Patients	EIS - Eisai Limited
O - Optometrist	D - Diabetes Assistance Program	EMD - EMD Serono Canada Inc.
	E - Exception status applies	GSK - GlaxoSmithKline Inc.
		PFI - Pfizer Canada Inc.
		SHI - Shire Canada Inc.