SCHEDULING STATUS

Schedule 4

ZAVESCA 100 mg

Hard capsules

Miglustat

Sugar free

Read all of this leaflet carefully before you start taking ZAVESCA

- Keep this leaflet. You may need to read it again.
- If you have any further questions, ask your doctor, pharmacist or nurse or other health care provider
- ZAVESCA has been prescribed for you personally and you should not share your medicine with other people. It may harm them even if their symptoms are the same as yours.

What is in this leaflet

1. What ZAVESCA is and what it is used for
2. What you need to know before you take ZAVESCA
3. How to take ZAVESCA
4. Possible side effects
5. How to store ZAVESCA
6. Contents of the pack and other information

1. What ZAVESCA is and what it is used for

ZAVESCA contains the active substance miglustat which belongs to a group of medicines that affect metabolism. It is used to treat two conditions:

- **ZAVESCA is used to treat mild to moderate type 1 Gaucher disease in adults**

In type 1 Gaucher disease, a substance called glucosylceramide is not removed from your body. It starts to build up in certain cells of the body's immune system. This can result in liver and spleen enlargement, changes in the blood, and bone disease.

The usual treatment for type 1 Gaucher disease is enzyme replacement therapy. ZAVESCA is only used when a patient is considered unsuitable for treatment with enzyme replacement therapy.

- **ZAVESCA is also used to treat progressive neurological symptoms in Niemann-Pick type C disease in adults and in children**.

If you have Niemann-Pick type C disease, fats such as glycosphingolipids build up in the cells of your brain. This can result in disturbances in neurological functions such as slow eye movements, balance, swallowing, and memory, and in seizures.

Zavesca works by inhibiting the enzyme called "glucosylceramide synthase" which is responsible for the first step in the synthesis of most glycosphingolipids

2. What you need to know before you take ZAVESCA

Do not take ZAVESCA:

- if you are hypersensitive (allergic) to miglustat, or any of the other ingredients of this medicine (listed in section 6).

Warnings and precautions

Talk to your doctor or pharmacist before taking ZAVESCA

- if you suffer from kidney disease
- if you suffer from liver disease

Your doctor will perform the following tests before treatment and during treatment with ZAVESCA:

- an examination to check the nerves in your arms and legs
- measurement of vitamin B12 levels
- monitoring growth if you are a child or adolescent with Niemann-Pick type C disease
- monitoring of blood platelet counts

The reason for these tests is that some patients have had tingling or numbness in the hands and feet, or a decrease in body weight, while taking ZAVESCA. The tests will help the doctor decide whether these effects are due to your disease or other existing conditions, or due to side effects of ZAVESCA (see section 4 for further details)

If you have diarrhoea, your doctor may ask you to change your diet to reduce your lactose and carbohydrate intake such as sucrose (cane sugar), or not to take ZAVESCA together with food, or to temporarily reduce your dose. In some cases the doctor may prescribe anti-diarrhoeal medicines such as loperamide. If your

diarrhoea does not respond to these measures, or if you have any other abdominal complaint, consult your doctor. In such case, your doctor may decide to conduct further investigations**.**

Male patients should use reliable birth control methods during their treatment with ZAVESCA, and for 3 months after finishing treatment.

Children and adolescents

Do not give ZAVESCA to children and adolescents (below 18 years old) with type 1 Gaucher disease because it is not known if it works in this disease.

Other medicines and ZAVESCA

Always tell your healthcare provider if you are taking any other medicine. (This includes all complementary or traditional medicines).

Tell your doctor if you are taking medicines containing imiglucerase, which are sometimes used at the same time as ZAVESCA. They may lower the amount of ZAVESCA in your body

Pregnancy and breastfeeding and fertility

You should not take ZAVESCA if you are pregnant or thinking of becoming pregnant. Your doctor can give you more information. You must use effective birth control while taking ZAVESCA. Do not breast-feed while you are taking ZAVESCA.

Male patients should use reliable birth control methods during their treatment with ZAVESCA, and for 3 months after finishing treatment.

If you are pregnant, breast feeding, think you may be pregnant or planning to have a baby, ask your doctor or pharmacist for advice before taking this medicine.

Driving and using machines

ZAVESCA may make you feel dizzy. Do not drive or use any tools or machines if you feel dizzy.

It is not always possible to predict to what extent ZAVESCA may interfere with the daily activities of a patient. Patients should ensure that they do not engage in the above activities until they are aware of the measure to which ZAVESCA affects them.

3. How to take ZAVESCA

Do not share medicines prescribed for you with any other person.

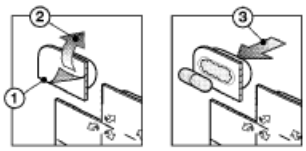
Always take ZAVESCA exactly as your doctor or pharmacist has told you. Check with you doctor or pharmacist if you are unsure.

• **For type 1 Gaucher disease**: For adults, the usual dose is one capsule (100 mg) three times a day (morning, afternoon and evening). This means a daily maximum of three capsules (300 mg).

• **For Niemann-Pick type C disease**: For adults and adolescents (over 12 years old), the usual dose is two capsules (200 mg) three times a day (morning, afternoon and evening). This means a daily maximum of six capsules (600 mg).
For children less than 12 years old, your doctor will adjust the dose for Niemann-Pick type C disease.

If you have a problem with your kidneys you may receive a lower starting dose. Your doctor may reduce your dose, e.g., to one capsule (100 mg) once or twice a day, if you suffer from diarrhoea when taking ZAVESCA (see section 4). Your doctor will tell you how long your treatment will last.

To remove the capsule:



1.Separate at perforations

2.Peel back paper at arrows

3.Push product through foil

ZAVESCA can be taken with or without food. You should swallow the whole capsule with a glass of water.

Your doctor will tell you how long your treatment with ZAVESCA will last. Do not stop treatment early because you need your medication. If you have the impression that the effect of ZAVESCA is too strong or too weak, tell your doctor or pharmacist.

If you take more ZAVESCA than you should

ZAVESCA has been used in clinical trials at doses ten times higher than the recommended dose: this caused decreases in white blood cells and other side effects similar to those described in section 4. In the event of overdosage, consult your doctor or pharmacist. If neither is available, contact the nearest hospital or poison centre.

If you forget to take ZAVESCA

Take the next capsule at the usual time. Do not take a double dose to make up for forgotten individual doses.

If you stop taking ZAVESCA

Don't stop taking ZAVESCA without talking to your doctor.

If you have any further questions on the use of this medicine, ask your doctor or pharmacist.

4. Possible side effects

ZAVESCA can have side effects

Not all side effects reported for ZAVESCA are included in this leaflet. Should your general health worsen or if you experience any untoward effects while taking ZAVESCA, please consult your healthcare provider for advice.

Most serious side effects:

Tell your doctor immediately if you experience the following:

- Some patients have had tingling or numbness in the hands and feet (seen commonly). They could be signs of peripheral neuropathy, due to side effects of ZAVESCA or they could be due to existing conditions. Your doctor will perform some tests before and during treatment with ZAVESCA to assess this (see section 2).

- If you get a slight tremor, usually trembling hands, seek medical advice from your doctor as soon as possible. The tremor often disappears without needing to stop the treatment. Sometimes your doctor will need to reduce the dose or stop ZAVESCA treatment to stop the tremor.

Frequent Side Effects:

- The most frequent side effects are diarrhoea, flatulence (wind), abdominal (stomach) pain, weight loss and decreased appetite.
- If you do lose some weight when you start treatment with ZAVESCA don't worry. People usually stop losing weight as treatment goes on.
- Other frequent side effects of treatment include headache, dizziness, paraesthesia (tingling or numbness), abnormal coordination, hypoaesthesia (reduced sensation to touch), dyspepsia (heartburn), nausea (feeling sick), constipation and vomiting, swelling or discomfort in the abdomen (stomach) and thrombocytopaenia (reduced levels of blood platelets).The neurological symptoms and thrombocytopaenia could be due to the underlying disease.

- Other possible side effects are muscular spasms or weakness, fatigue, chills and malaise, depression, difficulty sleeping, forgetfulness, and less libido.

Most patients get one or more of these side effects, usually at the start of treatment or at intervals during treatment. Most cases are mild and disappear quite quickly. If any of these side effects cause problems, consult your doctor. He or she may reduce the dose of ZAVESCA or recommend other medicines to help control side effects.

If you notice any side effects not mentioned in this leaflet, please inform your doctor or pharmacist.

Reporting of side effects

If you get any side effects, talk to your doctor or pharmacist. This includes any possible side effects not listed in this leaflet.

You can also report side effects to SAHPRA via “6.04 Adverse Drug Reaction Reporting Form”, found online under SAHPRA’s publications:

https://www.sahpra.org.za/Publications/Index/8.

By reporting side effects, you can help provide more information on the safety of ZAVESCA.

5. How to store ZAVESCA

Store all medicines out of reach of children

Do not take this medicine after the expiry date which is stated on the carton after 'EXP'. The expiry date refers to the last day of that month.

Store at or below 30°C.

Keep the tablets in the original packaging until required for use.

Do not store in a bathroom.

Return all unused medicine to your pharmacist.

Do not throw away any medicines via wastewater or household waste.

6. Contents of the pack and other information

What ZAVESCA contains

The active substance is miglustat 100 mg.

The other ingredients are:

Sodium starch glycollate,

Povidone (K30),

Magnesium stearate.

Gelatine,

Titanium dioxide (E171),

Black iron oxide (E172),

Shellac

What ZAVESCA looks like and contents of the pack

Opaque capsules with a white body and a white cap containing a fine white to off white powder. The cap is marked with 'OGT 918' and the body is marked with '100'.

Blister pack comprises of thermoformed Aclar UltRx 2000 adhesive laminated to opaque white PVC film pockets lidded with a child resistant plain hard–tempered aluminium foil, coated with heat seal lacquer and printed paper side.

Pack size: 21 capsules per blister strip and 4 blister strips per outer carton.

HOLDER OF CERTIFICATE OF REGISTRATION

JANSSEN PHARMACEUTICA (PTY) LTD

(Reg. No. 1980/011122/07)

2 Medical Road, Halfway House,

Midrand 1685, South Africa

MedInfoZA@its.jnj.com

Registration/Application number(s)

49/32.16/0152

Access to the corresponding Professional Information

Included in the carton, accompanying this patient information leaflet.