



Ambrisentan

This information should be read in conjunction with any patient information leaflet provided by the manufacturer. However, please note that this information sheet explains about the use of ambrisentan in children and young people so may differ from the manufacturer's information.

This information sheet explains about ambrisentan, how it is given and some of the possible side effects. Each person reacts differently to medicines, so your child will not necessarily suffer every side effect mentioned. If you are concerned about any of these side effects, please ring one of the contact numbers on this information sheet and ask for your doctor, nurse or pharmacist.

What is ambrisentan?

Ambrisentan belongs to a group of medicines called 'endothelin receptor antagonists'. It is prescribed at Great Ormond Street Hospital (GOSH) to treat pulmonary hypertension (high blood pressure in the blood vessels in the lungs). Ambrisentan acts against a substance called endothelin, which causes blood vessels to narrow and makes it more difficult for blood to flow through them. Ambrisentan relaxes the blood vessels so that blood can flow through the vessels more easily.

The use of ambrisentan in children is not currently licensed in the UK. Medicines are often used outside of their license (off-label) in children because clinical trial data is not available for a specific use. Prescribing medicines 'off label' is not necessarily hazardous but should be explained and agreed before use.

How is it given?

Ambrisentan is available as 5mg tablets under the brand name Volibris®. Unfortunately, no liquid preparation is available yet. Ambrisentan is given by mouth as a tablet once a day. It does not work immediately so it may take a month or two before you notice any improvement in your child's symptoms.

Ambrisentan is very expensive, so if only half a tablet is required per dose, the remaining half tablet should be saved for the next dose. Your doctor, nurse or pharmacist will have explained to you how to give the dose before you leave hospital.

Pregnant women and women of child-bearing potential who may need to cut tablets should avoid exposure to ambrisentan dust. The risk of exposure to ambrisentan tablet dust when cutting ambrisentan tablets is likely to be low. However, if you are concerned, then disposable gloves and a face mask should be used.

Who should not take ambrisentan?

People with the following conditions should discuss taking this medicine with their doctor.

- Hypersensitivity to ambrisentan or any of its ingredients, including soya and lactose
- Existing liver problems
- Scarring of the lungs (idiopathic pulmonary fibrosis)
- Pregnant, could be pregnant, planning to become pregnant or breastfeeding

What are the side effects of ambrisentan?

- Ambrisentan can cause anaemia and liver damage. Your child will need regular blood tests to check for anaemia and to check if their liver is functioning properly.
- Blood tests are required before starting ambrisentan and then every month during treatment. More frequent tests may be needed when the dose is altered.

Signs that the liver may not be working properly include nausea, vomiting, fever, pain in the stomach, jaundice (yellowing of skin or the whites of eyes), dark-coloured urine, itching, unusual tiredness or exhaustion, flu-like syndrome (joint and muscle pain, with fever). If your child displays any of these symptoms, tell your doctor immediately.

If any of these side effects are severe or carry on for a long time, please tell your doctor

- Upset stomach or abdominal pain
- Headache
- Dizziness
- Nasal congestion or sinus pain
- Flushing
- Low blood pressure (light-headedness)
- Heart palpitations
- Swelling or fluid retention
- Feeling tired
- Worsening shortness of breath

Some people develop an allergic reaction to ambrisentan, which may be mild or severe. Signs of a mild reaction include skin rashes and itching, high temperature, shivering, redness of the face, a feeling of dizziness or a headache. If you see any of these signs, please report them to a doctor or nurse.

Signs of a severe allergic reaction include any of the above, as well as shortness of breath or chest pain. If you are in hospital and your child shows signs of a severe allergic reaction, call a doctor or nurse immediately. If you are at home, call an ambulance immediately. If your child has a severe reaction to ambrisentan, subsequent treatment will be changed.

Fertility: Ambrisentan may lower sperm counts. In male children, it is possible that ambrisentan may have a long-term impact on fertility. Talk to your child's doctor if you have any questions or concerns about this.

Pregnancy: Ambrisentan may be harmful to unborn babies if taken during pregnancy. Females aged 12 years or older should use barrier contraception if sexually active. Please discuss contraception with the doctor. Monthly pregnancy tests are also recommended in female patients who are sexually active.

Ambrisentan and other medicines

Some medicines can react with ambrisentan, altering how well it works. Always check with your doctor or pharmacist before giving your child any other medicine, including medicines on prescription from your family doctor (GP), medicines bought from a pharmacy (chemist) or any herbal or complementary medicines

The following medicines may be taken with ambrisentan on the advice of your doctor but the dosages might need to be altered:

- Ciclosporin
- Rifampicin

Important information

- As this is a specialist treatment, your child's medicine will be supplied by a homecare service. You will need to contact the homecare company at least 14 days before your supply runs out to order further supplies.
- If you have any queries regarding deliveries or supply, please contact the homecare company, or the Pulmonary Hypertension team.
- Keep medicines in a safe place where children cannot reach them.
- Keep the tablets their original packaging, in a cool, dry place away from direct sunlight and heat.
- If you forget to give your child a dose and it is within a few hours of when the dose was due, give it as soon as you remember. Otherwise, do not give this dose but give the next dose when it is due. Do not give a double dose.
- If your child vomits straight after taking the dose, inform your local doctor or nurse, as your child may need to take another one.
- Your child should not stop taking ambrisentan suddenly. Your doctor may need to reduce the dose gradually over a period of time.
- If your doctor decides to stop treatment or the tablets pass their expiry date, return any unused tablets to the homecare company or alternatively your local hospital or pharmacy. Do not flush them down the toilet or throw them away.

Useful numbers

GOSH switchboard: 020 7405 9200

Pharmacy medicines information: 020 7829 8608 (Monday to Friday from 9am to 5pm)

Pulmonary hypertension team: extension 1007 or 1005 via switchboard