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Mastocytosis and Anaesthesia Advice for patients

What is mastocytosis?

Mastocytosis is a very rare disorder affecting mast cells. In people who do not have mastocytosis, mast cells are triggered during allergy attacks. They release substances that cause the symptoms of allergy. These substances are stored in granules within the mast cells and are released into the body during an allergy attack. This is called 'mast cell degranulation'.

In people with mastocytosis, mast cells are triggered much more easily. This causes severe allergy-type symptoms, such as low blood pressure and swelling, in response to simple everyday events. For example, mast cells can be triggered by stress, exercise, drugs, food or alcohol, as well as by environmental factors such as excessive heat or cold.

There are several different types of mastocytosis and it can affect adults and children.

How is mastocytosis diagnosed?

Mastocytosis is usually diagnosed by hospital specialists such as haematologists (doctors who specialize in blood disorders) or dermatologists (skin specialists). **Patients with mastocytosis should be seen by an allergy specialist doctor so that they can be assessed for the risk of severe reactions and given training in how to manage attacks.** An important blood test is Mast Cell Tryptase. This is usually high in mastocytosis patients. The test will have been done at diagnosis and doesn't need repeating before surgery.

Is anaesthesia safe in patients with mastocytosis?

Worldwide, there are very few cases reported of serious problems occurring during anaesthesia in patients with mastocytosis.

In patients with mastocytosis an attack could be triggered by the operation itself (the physical trauma to the body) or by anaesthetic drugs. It could also be triggered by skin antiseptics or by latex gloves used during surgery. Fortunately, these incidents are very rare. Your allergy specialist can assess you for the risk of any of these triggers.

will the anaesthetist know about mastocytosis?

All anaesthetists in the UK are medically qualified and they are trained specifically to recognize and treat anaphylaxis (core curriculum, The Royal College of Anaesthetists). In addition, there are regional specialized clinics where patients who have had allergic reactions to anaesthetics are seen. We recommend that the patient can either be referred to one of these clinics or the anaesthetist can seek advice from the special clinic. A list of these clinics is available on the Association of Anaesthetists of Great Britain and Ireland website (www.aagbi.org/safety/allergies-and-anaphylaxis).

What precautions should be taken before anaesthesia?

General precautions

This information about mastocytosis is available to all anaesthetists via the Royal College of Anaesthetists website.

Many people with mastocytosis wear a Hazard Alert bracelet (e.g. MedicAlert). This alerts the anaesthetist in the event of emergency surgery, for example if the person is too unwell to be able to speak after head injury.

Patients with mastocytosis should be given the opportunity to discuss their anaesthetic with a Consultant anaesthetist. Most hospitals have pre-operative assessment outpatient clinics and your surgeon should refer you to the appropriate anaesthetic clinic before planned surgery. We recommend that the anaesthetist who sees you in the pre-operative clinic should obtain advice from the nearest allergy/immunology clinic specializing in anaesthetic reactions.

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Every effort should be made to locate records of any previous anaesthetics, although sometimes previous anaesthetic records may be unavailable. Patients should avoid any known triggering factors as well as any drugs that are known to trigger mast cells. Previous uneventful administration of a drug does not guarantee that a later anaesthetic including the same drug will be trouble-free and your anaesthetist will be on their guard to pick up any adverse reaction immediately if it occurs.

Stress and sudden changes in temperature are potential triggers in mastocytosis. Operating theatre staff recognize that avoiding stress and excessive temperature changes is important for all patients. A mild sedative pre medication may be given on the morning of surgery. In children, being given gas may be less stressful than having an injection to go off to sleep. This approach may be appropriate for children who have previously reacted to the local anaesthetic creams often used to numb the skin before an injection.

During abdominal surgery the surgeon may need to move the bowel and this has been reported to trigger mast cell degranulation.

Anaesthetic drugs

Many patients with mastocytosis have been anaesthetized uneventfully. Therefore it is possible to make recommendations about which anaesthetic drugs are likely to be safe. A large number of different anaesthetic drugs are available. Unfortunately, much of the information available about drug safety on the internet is not evidence based and may be over-generalised.

Patients should avoid any drug which has caused worsening of their symptoms in the past. Some drugs are known to cause degranulation of mast cells, even in patients without mastocytosis, and these drugs should be avoided (see below). It is wise for intravenous drugs to be administered slowly and in the minimum dose required.

Analgesics (drugs given for pain relief)

Some pain killers are more likely to trigger a reaction, for example, codeine, morphine, pethidine and buprenorphine. On the other hand, Fentanyl is a versatile pain killer, which is less likely to trigger a reaction. Fentanyl can be given into a vein and is also available as a skin patch for more prolonged use. Post-operative pain can be treated with a patient-controlled infusion of fentanyl into a vein, or with a fentanyl skin patch. Remifentanil is another analgesic drug, which does not cause mast cell degranulation and is likely to be safe in mastocytosis patients, but this drug can only be given during anaesthesia because it is very potent. Paracetamol has been used safely in mastocytosis. Aspirin and ibuprofen (NSAIDs) have also been given safely but some mastocytosis patients find that their symptoms are triggered by NSAIDs, in which case all NSAIDs should be avoided.

Muscle relaxant drugs (drugs which relax the muscles, making some types of surgery easier, espeically operations in the abdomen or in the chest)

Atracurium and mivacurium should be avoided because they cause mast cell degranulation. Rocuronium, vecuronium and cisatracurium are suitable alternatives. Succinylcholine (suxamethonium) is unlikely to cause non-allergic mast cell degranulation but it remains the commonest cause of allergic anaphylaxis during anaesthesia. Succinylcholine should be avoided if possible because, if allergic anaphylaxis occurs in a patient with mastocytosis, the reaction is likely to be more severe than in patients who do not have mastocytosis.

Induction agents (drugs given at the beginning of the anaesthetic to cause unconsciousness)

Propofol is the most commonly-used induction drug and it is suitable in patients with mastocytosis. It can be used as an induction agent or as a continuous infusion to maintain anaesthesia throughout surgery.

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Inhalational agents (anaesthetic gases which keep patients unconscious until after the operation has finished)

These do not cause mast cell degranulation and are therefore safe. Examples are sevoflurane, isoflurane and desflurane.

Sedatives

There is no particular reason to avoid benzodiazepines such as diazepam, temazepam and midazolam.

Local anaesthetics

There is no evidence that local anaesthetics of the common amide type should be avoided (lidocaine, bupivacaine, levobupivacaine, ropivacaine, prilocaine) Amethocaine, an ester local anaesthetic found in Ametop local anaesthetic cream, should probably be avoided because it is known to cause localised skin reactions in some patients. EMLA cream (contains lidocaine and prilocaine) is a suitable alternative. There is no reason to avoid epidural or spinal anaesthesia, or local anaesthetic nerve blocks.

Anti-sickness drugs (also called anti-emetics)

These drugs have been given safely. Some of the antiemetic drugs also have antihistamine effects, which may be beneficial.

Skin antiseptics (used to clean the skin)

Skin antiseptics are necessary before surgery to reduce the possibility of infection. The two commonly used skin antiseptics are povidone iodine (e.g. Betadine) and chlorhexidine (e.g. Hibitane, ChloraPrep). Allergic reactions are more common with chlorhexidine than with povidone iodine. If there is a concern that there may have been a previous reaction to iodine or any skin antiseptic, further investigations (e.g. skin testing) should be performed by an allergy specialist.

Are any special tests needed before having an anaesthestic?

Assuming that the diagnosis of mastocytosis has already been made, no special tests are required. However, it is recommended that patients with mastocytosis who are expecting to undergo surgery should be reviewed beforehand by an allergy specialist who can discuss the appropriate management with the anaesthetist and the GP before surgery. Mastocytosis patients have skin which often reacts to pressure and touch with some swelling. F or this reason, the usual skin prick testing done in allergy clinics may give unreliable results.

Should patients with mastocytosis receive antihistamines before their anaesthetic?

Patients with mastocytosis have been anaesthetised uneventfully with or without being given anti-histamine drugs (H1 receptor antagonists, e.g. chlorphenamine or Piriton) beforehand. However, as a general principle it is appropriate for mastocytosis patients to receive antihistamines before the start of anaesthesia.

Should patients take sodium cromoglicate before their operation?

Sodium cromoglicate has been shown to be effective for some of the effects of mastocytosis on the gut (for example, diarrhoea and abdominal pain). Cromoglicate does not enter the bloodstream very effectively and may not be very effective in preventing low blood pressure. If you are routinely taking cromoglicate by mouth you could continue taking it up to the time (night before) of the operation. If you are not already taking cromoglicate, there is no evidence that you should start taking it before an operation.

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Should patients start steroids before an operation?

There is no evidence to show that this is effective. Even short term steroids can cause problems like poor wound healing. Steroids take several hours to work; so if they are going to be given, they need to administered several hours before the operation.

Should patients change their normal medication before anaesthesia?

Expert medical advice should be sought before changing any long-standing medication. Patients should continue taking drugs that they normally take to reduce their symptoms.

Beta-blocking drugs (beta adrenergic receptor antagonists) could potentially reduce the effectiveness of epinephrine (adrenaline) in restoring normal blood pressure if a patient experiences significant mast cell degranulation during anaesthesia. Nevertheless, the effects of usual doses of beta-blocking drugs can be overcome by increasing the dose of epinephrine (adrenaline) in the event of anaphylaxis.

If a patient with mastocytosis is receiving a beta-blocking drug it may be appropriate to change to an alternative drug several weeks before non-urgent surgery. However, in patients who have coronary artery disease beta-blocking drugs are valuable in protecting against a heart attack in the perioperative period and beta-blocking drugs should probably be continued in these patients if they have not caused problems previously. If the beta-blocking drug is being taken for high blood pressure, it is probably appropriate to change to a different type of drug a few weeks before surgery.

ACE inhibitors are another type of drug used to reduce blood pressure (the names of ACE inhibitor drugs usually end in ' ...pril'; for example enalapril, perindopril, lisinopril).

These drugs make the effects of mast cell degranulation worse and they should be stopped a few weeks before planned surgery, after taking advice from the GP and an allergy specialist.

Angiotensin receptor antagonists are also used to lower blood pressure (the names of these drugs usually end in '...sartan'; for example losartan, candesartan). These are likely to be safer and you are more likely to be advised to continue them.

How do you monitor patients for reactions during anaesthetics?

Patients in operating theatres are routinely connected to monitors such as blood pressure, pulse, oxygen level and ECG (heart rhythm) and these will reliably and instantly pick up signs of reactions. The anaesthetist will be with you throughout the operation, monitoring you carefully.

What drugs will be given to treat an attack during an anaesthetic?

Just as for a conscious patient, the most effective drug is adrenaline, which is available in all operating theatres.

Should x-ray contrast be avoided?

Radiologists sometimes need to inject a drug (contrast medium) before taking X-rays. Contrast media can sometimes trigger mast cells. It may be possible to avoid contrast media in some circumstances. This is best discussed with the radiologist beforehand.

Should an hdu or icu bed be available?

To ensure the best post operative monitoring, we recommend that an HDU bed is booked for the first night after surgery. This is to look out for delayed reactions and any problems caused by pain killers after the operation. Daycase surgery is probably not appropriate for this reason.

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Where do i find an allergy specialist?

A list of allergy specialists in the UK can be found on the website of the British Society for Allergy and Clinical Immunology bsaci.org (www.bsaci.org/find-a-clinic/index.htm) and also on the website of the Association of Anaesthetists of Great Britain and Ireland (www.aagbi.org/safety/allergies-and-anaphylaxis).

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