**Cutaneous mastocytosis**

This information sheet from Great Ormond Street Hospital (GOSH) explains the causes, symptoms and treatment of cutaneous mastocytosis and where to get help.

**What is cutaneous mastocytosis?**

Cutaneous mastocytosis is a condition characterised by increased numbers of mast cells in the skin. Mast cells are part of the immune system. As a result of certain stimuli, including parasites and insect bites, mast cells release a number of chemicals, including histamine. Histamine makes blood vessels expand and can make soft tissue swell. Cutaneous mastocytosis is a relatively rare condition.

**What causes cutaneous mastocytosis?**

Cutaneous mastocytosis is caused by genetic mutation. Human beings have about 30,000 to 40,000 different genes. Inevitably some of these genes are faulty, which can have a knock on effect on the body’s processes and functions. The gene mutation for mastocytosis increases production of mast cells. It usually occurs as a sporadic or isolated change – it is not usually passed on from parent to child.

**What are the signs and symptoms of cutaneous mastocytosis?**

Cutaneous mastocytosis usually presents as lesions on the skin. These can become raised and red and even blister if rubbed. The lesions may appear brown, leading to the label ‘urticaria pigmentosa’.

The increased production of mast cells by the body increases the risk of allergic type reactions.

**How is cutaneous mastocytosis diagnosed?**

Cutaneous mastocytosis can be diagnosed using physical examination – one specific sign is that the skin lesions become red, itchy, and sometimes blister, if rubbed gently. Occasionally a skin biopsy may be suggested – a small sample of skin is taken and examined under a microscope to see if there are more mast cells present than expected.

**How is cutaneous mastocytosis treated?**

Cutaneous mastocytosis often requires no treatment, but if itching and blistering are troublesome antihistamines may be prescribed. As mastocytosis may increase the risk of an anaphylactic reaction, an injector pen containing adrenaline may be supplied. Some children with extensive involvement may be advised to wear a Medicalert bracelet or similar notification jewellery so that healthcare staff are aware of the diagnosis in case of emergency.

Rarely, other forms of treatment with other medicines or ultraviolet light might be suggested. Certain substances, medicines and physical factors can trigger reactions so these should be avoided where possible. These include:

- Alcohol
- Medicines including atropine, dextran, aspirin and non-steroidal anti-inflammatory drugs (NSAIDS), certain antibiotics, opioids, thiamine, quinine, gallamine, procaine, some radiographic dyes,
polymyxin B, amphotericin B, scopolamine, decamethonium, reserpine and tubocurarine.

- Bee and wasp stings and snake venom
- Extremes of temperature, sunlight or exercise

What happens next?
For most people, the condition improves by puberty.

Further information and support
The UK Mastocytosis Support Group can offer advice and support – visit their website at www.ukmasto.co.uk.