The XLP Research Trust
60 Winchester Road
Romsey, SO51 8JA
United Kingdom

+44 (0)1794 521077
info@xlpresearchtrust.org
www.xlpresearchtrust.org

Registered Charity No 1111075

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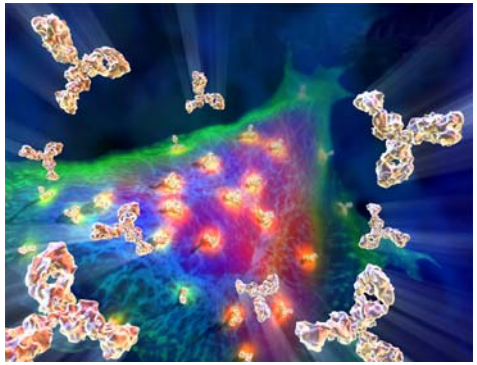
X-Linked Lymphoproliferative Syndrome

Immunoglobulin Replacement



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Introduction

One of the symptoms of XLP is hypogammaglobulinemia which means that there is a lack of B-lymphocytes and a resulting low level of immunoglobulins (antibodies) in the blood. This means that boys with XLP can be much more susceptible to infection and disease due to this lack of antibodies.

Replacement immunoglobulins, or IgG replacement, is a preparation of purified natural blood plasma components which when infused will boost the amount of antibodies in the XLP boys blood helping them to fight infection successfully. IgG replacement gives protection against measles, chickenpox, and most viral and bacterial infections. As a result children using IgG replacement do not normally need routine immunisations although this should be confirmed with your doctor. This level of antibodies needs to be topped-up every so often to maintain the protection.

IgG replacement is hence very important for a boys diagnosed with XLP but is **not** a cure of the underlying condition.

How is it given?

The most common way of giving immunoglobulin is by infusion through a cannula into a vein (intravenously or IV), but it can also be given by rapid subcutaneous infusions (SC) under the skin. Specialist centres are using the subcutaneous route increasingly and new products are being licensed specifically for this.

Intravenous immunoglobulin (IVIG)

Administering IVIG is dependent on having good venous access. Infusions are normally administered every three weeks to give good replacement blood IgG levels. All infusions should be started slowly and gradually increased over 30 to 60 minutes to the maximum rate. The dose of IVIG varies according to the patients weight. IVIG can be administered at most hospitals. Infusions normally take between three and four hours to complete and usually over one day.

Subcutaneous immunoglobulin (SCIG)

Subcutaneous immunoglobulin is becoming increasingly popular as a safe and effective way of administering this treatment. It is ideal for patients who have poor veins, and for small children who need smaller doses. It is easy to administer and therefore once patients are stable on treatment, families can be taught how to give the infusions at home reducing the need for regular hospital visits. Two infusions are given simultaneously through fine butterfly needles under the skin into the abdomen or thighs. Small portable infusion pumps are used to administer the infusions and they take 45 to 90 minutes depending on the amount of treatment given. The amount of fluid a child receives in each site depends on the dose and how much the skin can stretch to absorb it. Most children receive their infusions every week. Babies and small children are usually given 10mls, but it is possible to give up to 30mls per site in older children.

What are the possible risks or side-effects?

People receiving IgG replacement may occasionally experience (during or after the infusion) a chill, headache, abdominal pain, fever, nausea, vomiting and joint pain – particularly low back pain. These symptoms are known as an 'infusion reaction' and will usually settle quickly. If they occur during the infusion, then the infusion may be stopped or slowed down.

Occasionally people may experience an allergic reaction when their blood pressure may drop (sometimes causing them to feel faint or light-headed) and they may develop itchy skin, swelling of the face and throat, and have difficulty breathing. Very occasionally IgG replacement can cause a rise in blood pressure. A nurse will monitor you during the infusion but please report any new symptoms during or after the infusion. These reactions occur only in a minority of patients. Rarely, people receiving IgG replacement may experience a rash or abnormalities in liver function (detected by blood tests), but these usually settle quickly. Other rare side-effects can also occur. All these infrequent side-effects can be treated.

All blood donors from whom IgG replacement is made are carefully screened. However, it is impossible to eliminate completely the risk of passing on infection, for example with an unknown virus.

However the benefits of this treatment in preventing life threatening infections for XLP affected boys outweigh the potential risk of infections or side effects.

Definitions

Lymphocyte is a type of white blood cell found in your blood and bone marrow.

B lymphocyte—is a specific type of lymphocyte. It represents 5 to 15% of circulating lymphocytes in the blood. These are the cell that make immunoglobulins to fight infections.

Antibodies are types of protein found in the blood. They are produced by B lymphocytes. Their function is to bind to substances in the body that are recognised as foreign or potentially dangerous substances such as bacteria and viruses. These proteins circulate in the blood stream to attack the antigen and render it harmless.

Immunoglobulin is a class of antibody. There are five classes of immunoglobulin; of these, immunoglobulin G (IgG) is the major immunoglobulin in human blood and it this class that IVIG replaces or tops up.