

Great Ormond Street Hospital for Children NHS Foundation Trust: Information for Families

Morphoea

This leaflet explains about morphoea, how it is treated and what to expect when you come to Great Ormond Street Hospital (GOSH) for treatment.

What is morphoea?

Morphoea is a rare skin condition where patches of skin become hardened and lose their normal texture, becoming shiny. Sometimes the bones and muscles underneath the patches of skin become affected as well. We do not know what causes morphoea, but it seems to affect slightly more females than males.

Morphoea is also sometimes called localised scleroderma. This means it is localised to the skin and is quite separate from the condition of systemic scleroderma which affects internal organs.

What are the symptoms?

The main symptom of morphoea is the distinctive patches of hardened and shiny skin. However, these patches can have knock-on effects depending on their position on the body. For example, if the patch is on the face, this can cause dental problems as the bone underneath the patch shrinks and becomes thinner. If the patch is on a limb, particularly over a joint, this can cause the joint to stiffen making it difficult to bend and straighten. It could also cause limb length discrepancy where one limb is shorter than the other.

How is morphoea diagnosed?

Morphoea is usually diagnosed by a clinical examination of the lesions by a dermatologist (doctor specialising in skin disorders). Sometimes a biopsy (skin sample taken using a local anaesthetic injection) is done but this is not always necessary.

What treatments are available?

The aim of treatment is to stop the disease getting any worse, which will prevent complications developing. It also aims to improve the texture of the skin, although the patches can never be completely cleared. Morphoea is usually treated with two types of medications: steroids and immunosuppressive medications. The steroids are used to put the disease into remission and the immunosuppressive medication keeps it under control so it does not come back.



Steroids

Two types of steroids are used: methylprednisolone and prednisolone.

Methylprednisolone is given at the start of treatment as an intravenous infusion. Your child will need to come to GOSH for two hospital admissions, each about three days long, one week apart. Methylprednisolone is usually tolerated well, but a few side effects have been reported. These commonly include high blood pressure and high sugar levels in the blood and urine. While your child is having the methylprednisolone infusion they are closely monitored for any side effects, which can be treated with medication or by stopping the infusion.

For further information, please see our Pulsed methylprednisolone treatment information sheet.

Prednisolone is started after the first hospital admission for the methylprednisolone infusion. Prednisolone is given as tablets either to be swallowed whole or dispersed, and should be taken every day. The dose of tablets is gradually reduced depending on how your child responds to treatment, and is usually stopped after a couple of months. Like methylprednisolone, prednisolone is usually tolerated well, and similar side effects have been reported. Other side effects like hyperactivity and an increased appetite have also been reported.

For more information about steroids, please see our *Short-term steroid* treatment leaflet.

Immunosuppressive medications

One type of immunosuppressive medication is used, called methotrexate, which works by damping down the immune system. While your child is taking methotrexate, he or she should also take a vitamin called folic acid, which prevents some side effects developing.

Methotrexate is started during the hospital admission for the methylprednisolone infusion. It is given once a week by mouth (orally) either as tablets, syrup or as an injection under the skin (subcutaneously). The consultant will discuss both options with you. Your child will usually start by taking the tablets or syrup, but if they cannot tolerate these, the injections can be given instead. Your child will need to continue to take methotrexate for a period of one to three years to make sure that the morphoea has completely 'burnt out' and is unlikely to return.

The most common side effects are nausea (feeling sick) and stomach upsets. Alterations in blood count or liver function tests are also possible, but these are generally mild and controlled by reducing the dose of methotrexate or stopping it altogether for a short period. Alcohol increases the risk of liver damage, so this should be avoided while taking methotrexate. Methotrexate can damage the unborn baby. If your daughter is 12 years old or older, we will ask her about her periods and any possibility that she could be pregnant. If your daughter is sexually active, she should use a reliable form of contraception.



Please read our leaflets Methotrexate for chronic inflammatory conditions and Special handling requirements for oral cytotoxics and cytotoxic immunosuppressant medications for further details.

Check ups

Your child will need to have regular check-ups with your family doctor (GP) or local paediatrician while receiving treatment for morphoea. While your child is taking prednisolone, they should have regular blood pressure and urine tests.

Your child will need to have regular blood tests while taking methotrexate. These blood tests check for the less common side effects mentioned above. You will be given a card to record these results. Please remember to bring it to your clinic appointment.

Immunisations and infectious diseases

As steroids and methotrexate affect your child's immune system, they should not be given any 'live' vaccines, like polio, MMR or BCG. The 'killed' or inactivated vaccine should be used instead. Your other children should be immunised according to the usual schedule but they should also receive the 'killed' or inactivated vaccine. If you have any questions about immunisations, please ask your doctor.

If your child has not had chicken pox and is in direct contact with a child who has chicken pox or develops it within 48 hours, you must contact your GP or local paediatrician immediately. Chicken pox can be more severe in children who are taking methotrexate or steroids. A blood test will be arranged to check your child's antibodies to chicken pox. Your child may need an injection to protect them. Your doctor or nurse will discuss this with you.

What is the outlook for children with morphoea?

If your child has developed complications associated with morphoea, like dental or joint problems, they may need to be referred to other specialists for treatment. The outlook for children with morphoea is generally good as it responds well to treatment. In most cases, the condition 'burns out' after a few years of treatment, but there is a small chance it could recur. Your child will receive long-term follow up treatment once the morphoea has 'burnt out' to check whether it is recurring and to restart treatment if this is the case.

Is there a support group?

The **RSA** - Raynaud's and Scleroderma Association – can offer support and information to anyone affected by morphoea. Call their helpline on 0800 917 2494 or visit their website at www.raynauds.org.uk

Compiled by the Birthmark Unit in collaboration with the Child and Family Information Group

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