

Information Leaflet for parents/carers

Acral Peeling Skin Syndrome



Acral Peeling Skin Syndrome

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We hope that this leaflet will help you to understand all about Acral Peeling Skin Syndrome.

What is Acral Peeling Skin Syndrome?

Acral Peeling Skin Syndrome (APSS) is a rare genetic skin fragility condition.

What are the symptoms?

The main symptom is painless peeling of the skin, mainly on the hands and feet. The word “acral” means “the extremities”. Children with APSS may also experience itching and reddening of the skin. The signs and symptoms of peeling skin usually appear soon after birth, but they may also develop later in life.

How is APSS diagnosed?

It is often confused with another skin condition called Epidermolysis Bullosa Simplex; APSS is diagnosed by taking a blood sample.

What causes APSS?

It is caused by a defective gene. Everybody carries two copies of every gene, one from each parent. APSS is an autosomal recessive condition. This means that a person with APSS has inherited a defective copy of the gene from both parents.

Mutations (changes) in a gene called TGM5 are the cause of most cases of APSS.

With APSS, the parents usually have one defective copy and one normal copy: they are said to be “carriers” and do not have APSS.

Parents who have a child with APSS have a one in four chance of any future children developing the condition. People with APSS are very unlikely to pass it on to their own children as they will only pass on one copy of the mutated gene.

How is APSS treated?

There is currently no cure for APSS, therefore treatment is centred on preventing skin damage and helping to reduce symptoms. Peeling of the skin is the main symptom but blistering can also occur. It is best to burst new blisters with a sterile needle to release the fluid and then apply dressings, as appropriate.

The condition is usually made worse by hot temperatures, high humidity and friction; therefore the dressings need to be light and not too tight.

The dressings that are most commonly used are silicone dressings, such as Mepilex Border Lite, and Mepilex. These dressings do not

stick to the skin and can help wounds heal and prevent any open wounds from getting infected. Dressings can be held in place with a sock, cotton bandage or tubular bandage such as Tubifast. The peeling usually heals fully without leaving scars. However try not to let your child scratch the affected areas as this may lead to scarring.

What else do I need to know and how can I help?

Due to blistering, your child may experience sore hands and feet. Choosing the right footwear is very important. Allow your child to wear footwear that is comfortable and doesn't cause blisters, for example, trainers, sandals, Crocs or slippers.

Allow your child to remove their shoes to let their feet to cool down, if necessary. It may be appropriate to keep a spare pair of shoes for your child to change into at nursery or school.

If your child's fingers are very sore, soft hand grips can be applied to pencils, paint brushes etc.

To prevent infection and to moisturise the skin an antibacterial soap substitute, such as Dermol 500, should be used when washing. Symptoms may be worse in the summer due to the temperature and humidity.

Looking after and sharing information about your child

We have a duty of care to help patients and families understand how information about them is kept and shared and we include the following information in all our patient leaflets:

Information is collected about your child relevant to their diagnosis, treatment and care. We store it in written records and electronically on computer. As a necessary part of that care and treatment we may have to share some of your information with other people and organisations who are either responsible or directly involved in your child's care. If you have any questions and/or do not want us to share that information with others, please talk to the people looking after your child or contact PALS (Patient Advice and Liaison Service) on 0121 333 8403/8611.

Please use this space to write down any notes or questions you might have.

Support Groups

EB team at Birmingham Children's Hospital

Tel: 0121 333 8224/8757

eb.team@bch.nhs.uk

www.bch.nhs.uk

EB patients' support group DEBRA

Tel: 01344 771961

debra@debra.org.uk

www.debra.org.uk

Health information and internet access is also available in the Family Health Information Centre at the hospital.

Tel: 0121 333 8505

Email: child.infoctr@bch.nhs.uk

Further Information

We hope this leaflet will help you to understand about your child's condition. This leaflet was produced using the most up to date evidence available. Further details are available upon request. If you feel you need any more information please speak to your child's dermatologist or specialist nurse who will try to answer any further questions you may have.

Dermatologist Tel:

Specialist Nurses Tel:

Community Nurses Tel:

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