UK National Screening Committee

# Frequently asked questions – Severe Combined Immunodeficiency (SCID)

**Last updated:** February 2013

## What is Severe Combined Immunodeficiency (SCID)?

Severe Combined Immunodeficiency (SCID) is the name for a group of very rare genetic disorders. There are more than 10 types of SCID. All are caused by abnormalities in a baby's DNA (the instructions inside each cell of the human body that control what the cells do). Babies born with SCID have weakened immune systems and are unable to defend themselves against germs and infections.

## What can happen to children affected by SCID?

Babies born with SCID easily contract infections which can be serious or even life threatening. These infections can harm the lungs (pneumonia), brain (meningitis) or blood. Babies with SCID are also vulnerable to ear and sinus infections, chronic diarrhoea, skin rashes and thrush infections.

Sometimes babies with SCID develop multiple infections at the same time. These infections generally appear in the first few months of a baby's life.

## Who is at risk and what is being done to prevent this?

Unless there is a family history of SCID it is difficult to know who is at risk for SCID. SCID is a rare condition and recent estimates suggest that about 1 in every 35,000 babies in the UK will be born with the condition each year. It's very important that SCID is diagnosed as soon as possible, because without quick treatment, babies easily contract infections and are unlikely to reach age two.

If a baby begins to easily contract infections and the doctor suspects he or she might have SCID, there are tests that can be done. These tests can identify common signs of SCID, like low levels of white blood cells. The doctor will assess the test results to help determine if a baby has SCID and, if so, which treatment options are best.

### Why do we not have screening in the UK?

Newborn screening for SCID is a very new development. There is currently no country in the world with an established SCID screening programme to identify babies at birth, before they develop any symptoms. Universal screening for SCID was approved in the USA in 2010, and some states are currently running screening programmes. Pilot studies are also on going in Sweden and Germany.

While the UK does not currently offer screening for SCID, the UK National Screening Committee (UK NSC) is currently looking at evidence to see whether a national screening programme should be introduced. The consultation closed on 14 January 2013 and the Committee is expected to make a recommendation in spring 2013 to the Government on whether or not there should be a programme for SCID.

The UK NSC will only approve a screening programme if the benefits outweigh the harms. Based on the evidence and the responses to the consultation, the UK NSC will consider several factors, which include:

- **The test.** This needs to be simple, safe and accurate so that it picks up as many babies as possible with SCID without wrongly identifying others without the condition.

- **The treatment.** The Committee will only introduce a national screening programme if there is an effective treatment for SCID. The Committee will not introduce a screening programme for SCID if there is not a safe and effective treatment available.

- **Who is affected.** Before a national screening programme is established, the Committee will need to know how many babies in the UK are affected each year. This figure will help the Committee decide if a national screening programme is the best way to identify SCID. Currently, it is difficult to know exactly how many babies in the UK are born with SCID.

## What treatment options are available for babies with SCID?

Currently, the most effective treatment available for SCID is a bone marrow transplant. This involves taking the bone marrow of a healthy person and injecting into a baby with SCID. When successful, it can rebuild a child's immune system. Research reports that this allows babies affected by SCID to fight infections and live a normal, healthy life.

When considering whether a national screening programme is appropriate for SCID, the UK NSC will consider all available evidence about how successful treatment is and what the risks or harms might be. Bone marrow transplants can carry life threatening risks for both the child and the donor and the Committee will try to balance these and other risks against the benefits when deciding if a national screening programme is appropriate for SCID.

## *For more information*

The UK National Screening Committee (UK NSC) reviews the evidence for screening for conditions against strict criteria, which you can find here:
https://www.gov.uk/government/publications/evidence-review-criteria-national-screening-programmes

Details of the review process can be found here:
https://www.gov.uk/government/publications/uk-nsc-evidence-review-process