What to expect at your annual review - sickle cell disease

What is an annual review?

Your specialist centre may offer an annual review- this is a longer appointment, once a year, that is different in purpose from your usual clinic visits.

Most appointments are about how you are now – how your last crisis went, your medications, how you have been feeling. An annual review steps back from all that. It is about the long term: the things that build up quietly over years, and what can be done to stop them.

Your annual review lasts about 40 to 45 minutes. That is deliberate. It is the one appointment in the year with enough time to talk about everything, not just your blood results.

Because it is longer, it is worth preparing for – and worth arranging time off work or childcare for, if you need to. Coming in rushed, or having to leave early, can make this appointment less useful.

Why does it exist?

People with sickle cell disease are living longer than ever before – which is genuinely good news, and it has changed what we need to worry about.

As people live longer with sickle cell disease, we have become aware of complications that arise later in life. They tend to accumulate silently. Nothing hurts, nothing feels wrong, and by the time something does, damage has often already been done.

The reason that matters is what follows from it: **if good prevention is put in place, and if you work with us in sticking to those plans, in most cases the chance of these complications arising can be reduced**. That is why we set aside a longer appointment once a year, rather than trying to fit this around a routine visit.

It is also worth saying plainly: an annual review is not just about crises. It is entirely possible to have had a good year – no admissions, no serious pain – and still have something worth finding at this appointment. Feeling well is not evidence that nothing is happening.

Who will I see?

Usually a doctor and a specialist nurse, together or one after the other. In some services a psychologist is also part of the team.

If you meet a psychologist, this is not because anyone thinks there is something wrong with you or that you are not coping. Living with sickle cell disease – the pain, the appointments, the medicines, the way it shapes decisions about work and family – is a lot to carry, and it is treated as a normal part of your care rather than an add-on for when things go badly.

The specialist nurse is often the person you will have most contact with during the year, and the person easiest to reach when something comes up. The annual review is a good opportunity to make sure you know how to contact them.

What gets talked about?

This is the part people do not expect, and it is the reason for the extra time. An annual review is a conversation, not just a set of tests.

Your crises. How often you are having crises will be discussed- both the ones that bring you in to hospital and the ones that you treat at home. This is so that your medical team can review any triggers that you think causes your crises, and whether anything can be done to reduce these.

Your medicines, honestly. Not just what you are prescribed – whether you are actually taking it, and what gets in the way. This is a review of whether the plan is working for you, not a test of whether you have been good. If you have stopped your penicillin, if your hydroxyurea makes you feel rough, if the routine has fallen apart, this is the appointment to say so. Nobody is going to tell you off. Taking a tablet every day for a problem you cannot feel is genuinely hard, and stopping is common. Your team can only help with a problem they know about.

Pregnancy and contraception. Whether you are thinking about a pregnancy now, might be one day, or want to make sure you do not have one yet. In sickle cell disease there is a lot worth planning in advance rather than sorting out afterwards – some medicines need changing before a pregnancy rather than during it, and pregnancy and your condition affect each other in ways worth understanding early. Contraception choices can also be affected by your condition. Whatever your situation, this is the appointment to raise it. If you want to talk about testing for a future child, our leaflet on preimplantation genetic diagnosis (PGD) covers the options.

Priapism - for men and boys. You will be asked about this every year. Not because anyone suspects a problem, but because it is asked of everyone, every time – in exactly the same way as asking about your chest or your joints.

Priapism is a painful erection that happens without arousal, or that will not go away. It is caused by sickled cells blocking the blood draining away from the penis. Short episodes that come and go – often waking you at night, lasting under an hour, settling on their own – are called stuttering priapism, and they are common.

Most men do not bring them up. That is completely understandable, and it is also the problem: it is very easy to decide brief episodes are not worth mentioning. But they are worth mentioning. They tend to recur, treatment can reduce them, and over time repeated episodes can damage the tissue and cause lasting difficulty with erections. That damage is largely preventable – but only if your team knows the episodes are happening.

So the question gets asked routinely, so that you do not have to find a way to raise it. If you would rather discuss it with a male member of the team, or with the specialist nurse rather than in the main appointment, say so – that can usually be arranged. What matters is that it gets said.

An erection lasting more than one hour is a medical emergency. Go to A&E – do not wait to see if it settles, and do not wait until morning. Tell them you have sickle cell disease and that this is priapism. Treated early it usually resolves; after about four hours, permanent damage becomes much more likely.

Your joints. You will be asked about pain in your hips and shoulders. Sickle cell disease can reduce the blood supply to the end of a bone, causing the bone to be damaged – this is called avascular necrosis, or AVN. The hip is the commonest site, then the shoulder.

The reason it is asked about directly is that AVN pain is easily mistaken for an ordinary crisis, or simply absorbed into the background of living with sickle cell disease. It tends to be a deeper, more constant ache that does not behave like your usual pain – often worse on weight-bearing, worse over months rather than days, and not settling between crises.

There is no routine scan for this. It is picked up from what you describe and from examining you, so describing it is what starts the process. If AVN is suspected an MRI can find it – and finding it early matters, because there is more that can be done for a joint at an early stage than a late one.

Work, education and money. Whether your condition is affecting your job, your studies, or your finances. Time off for admissions and appointments, tiredness affecting your work or exams, and the practical business of benefits and support all come up here. Your team can advise and refer, and CAN can help too – please ask. People often assume this is not what the appointment is for. It is.

Staying well generally. Diet, exercise, alcohol, smoking, vaccinations, hydration, and travel plans. Not lecturing – these things genuinely interact with your condition, and there is time to discuss them properly.

Anything you have been meaning to ask. The things too small for a busy clinic and too persistent to forget. Write them down and bring them.

What gets checked?

Preventing serious infection. People with sickle cell disease have a spleen that does not work properly, even though it is still there. The spleen is critical for fighting certain infections, which is why two things matter:

Antibiotics. Daily penicillin – or an alternative if you are allergic – prevents very severe infections. In children these are life-saving, and the recommendation is that they continue into adulthood.

Vaccinations. The recommended schedule is a yearly flu vaccine, a full course of hepatitis B with a booster if needed, Hib once in a lifetime, meningococcal vaccine, pneumococcal every five years, and COVID.

Your eyes Sickle cell disease can cause bleeding from the small blood vessels at the back of the eye, which is why a yearly appointment with an optician or eye specialist matters. Changes can be found and treated before your vision is affected – you will not notice the early stages yourself.

Your kidneys. Kidney damage starts with protein appearing in the urine, long before you would feel anything. If it starts, there is medication that can slow it down. A urine sample is taken each year.

Your heart. An echocardiogram – an ultrasound of the heart – usually every three to five years.

Bone health. Sickle cell disease makes osteoporosis more likely – thinning of the bones – which can be prevented with calcium, vitamin D and exercise. Your vitamin D level is checked and replacement given if needed.

Your transfusions, if you have them. How often, and your red cell antibodies. Having your antibody list recorded matters enormously if you are ever transfused somewhere new.

Your history. Complications, surgery, and any past treatments.

An examination. Weight, oxygen levels, pulse, blood pressure, and an examination of your chest, heart and abdomen.

Why so many tests when I feel fine?

Because every one of them is looking for something that does not announce itself.

Kidney damage shows in a urine test long before you would notice anything. Damage at the back of the eye can be well advanced before your vision changes. Osteoporosis causes no symptoms until a bone breaks. Heart changes are found on a scan, not by feel.

Two of them are silent for a different reason: they are not asked about, or not mentioned. Repeated short episodes of priapism damage tissue quietly over years, and most men never raise them. Early avascular necrosis gets absorbed into the background of sickle cell pain and put down to a crisis. Neither shows up on any of the routine tests – they are found by asking, and answered by telling.

This is what "accumulating silently" means. The point of the annual review is to find these things while they can still be stopped or slowed – and that stage is, by definition, the stage where you feel completely well.

What happens afterwards?

You will receive a copy of the letter summarising what was discussed, your results, and the plan. Keep it.

That letter is the single most useful document you can have. If you are ever treated somewhere new – on holiday, at university, in an emergency, after a house move – it tells a team who has never met you what your diagnosis is, what antibodies you have, what you take, and who looks after you. A photograph of it on your phone takes a minute and can save hours at a difficult moment. This matters particularly in sickle cell disease, where you may arrive somewhere in a crisis and not be in a position to explain any of it.

If a follow-up test, scan or referral is arranged and you have not heard within the timeframe you were given, chase it. Things do occasionally get lost, and it is easier to ask than to find out a year later.

You do not have to wait for your annual review to raise something. If something changes or worries you before then, contact your team.

What if you don't think you are getting annual reviews?

Not all teams carry out annual reviews in the same way, and sometimes patients don't know that what they thought was a regular follow up appointment is actually the annual review. All teams have to carry out an annual review on every patient and report the results to the NHR (National Haemoglobinopathy Registry). See our leaflet about how haemoglobinopathy services are organized in England. You can ask your team to let you know when your last and next annual reviews are booked.

When to seek medical advice

Do not wait for your annual review if you notice:

- Fever – this always needs urgent attention
- Chest pain or difficulty breathing – seek help immediately
- An erection lasting more than one hour – go to A&E immediately
- Any change in your vision
- Increasing tiredness or breathlessness
- Pain in a hip or shoulder that is not part of a crisis, or that keeps coming back
- Swelling of the legs, or a change in how much urine you are passing
- New or worsening pain that is different from your usual pattern
- Any side effect making it hard to take your medication

Can we help?

CAN (congenital anaemia network) is a charity specifically for patients with inherited anaemias. It was started by a group of patients and doctors, working together to tackle some of the problems of having an inherited anaemia – the lack of information and the loneliness of not knowing anyone else with your condition. We run events for patients to meet each other, as well as provide some additional information, support and services for patients who need that extra bit of help.
If you want to know more visit www.togetherwecan.uk or email us on info@togetherwecan.uk

Do you know your care should involve a specialist centre for inherited anaemias? These conditions are rare enough that few doctors see many of them and all patients with rare inherited anaemias should be under the care of a haemoglobinopathy specialist. Please read our 'How red cell services are organised in England' information sheet. If you don't know who your specialist service should be, please get in touch with us so we can help you find out.