RCPCH The Paeds Round

The Secrets of Sickle Cell

Transcript of podcast –January 2026

[Music starts]

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Emma: You need to be a sickle cell champion to these children.

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Emma: I am very, very excited today, in November, in this horrible cold weather, to have a guest all the way from London to come and talk to us about one of the things that I think is really, really important and not talked about often enough, which is sickle cell disease. So, it's a huge welcome from me to you, Subarna, to come here and talk on the Paeds Round.

Subarna: Thank you. I greatly appreciate you asking me to come.

Emma: And just as a quick introduction, Subarna is fantastic expert. She is a paediatric haematologist. She works in London and has been working with sickle cell and Thalassemia for over 20 years and is the chair of the Hemoglobinopathy Reference group for NHS, England. So, who's better to talk about this than you?

You have nothing to say after that introduction! Okay, well, one of the things that I think is really useful for everybody listening is just to give us a little round up of the current sickle cell guidelines and the state of services for children with sickle cell in the UK. One of the things that strikes me is actually the NICE guidelines, so are actually quite old, so they're from 2012, so is that something that you think that we should think about, and what has happened since then?

Subarna: Yes, so the NICE guidelines specifically looks at the management of pain in sickle cell disease in the emergency department. And as you say quite rightly, it was published in 2012 and we haven't really had any updates since then. So that's not for the want of trying, though. In 2023 we went back with NICE to look at whether there was any new and emerging evidence to support or to change the guidance. But actually, what we

realised was that there wasn't any. So, we decided as a group of experts to kind of leave the NICE guidelines as they were.

And the essence of the guideline is really the time criticalness, for want of a better word, of providing pain relief to children and adults or anybody with sickle cell when they present to the emergency department. And this has been the cornerstone of sickle cell care in the UK, really, we as sickle cell care providers really feel it's really important that this is provided in a timely manner, pain relief. And also, if you look at emergency departments themselves, they too, not just for sickle cell, but they have very stringent guidance themselves for providing pain relief to anybody that comes with pain. In fact, theirs is a little bit more stringent than sickle cell, so, there's, I think is 15 minutes.

But the sickle cell guidance goes a little bit further than the first dose of analgesia, which ideally should be within 30 minutes. What it does say is that the provider, the nurse or the doctor looking after that patient, should then return to the patient every 30 minutes to check the response of the pain relief.

The other thing that is very important and slightly different from all other sort of pain approaches in ET is that one thing that nice guidance was very clear about was that you should treat sickle cell disease pain as an emergency. And I think that is important to underscore, because patients are often and I'm talking about really young children here, three-four-year-olds will often be able to be able to somehow regulate their own sense of pain in a way that they don't have any external visible reaction to the pain. So, and this is such a human thing that when we see somebody in pain, we then go back within ourselves, to dig into ourselves, to see, you know, what is my reaction to pain? So, people might experience trauma pain, people might experience surgical pain, people might experience birth, labour pain, etc. But for people who haven't experienced sickle pain, it's very difficult. And you know, for children who have experienced pain all their lives, and I'm hoping, and I will talk about that later, I'm hoping with newer, better treatments, we are trying to avoid that now, but for children who experience pain all their lives, are able to somehow kind of distract themselves from the pain. So often they are withdrawn, they are absolutely still. They will often try and concentrate, maybe on their phone, on a game, or do something. And this is their own way of coping with the pain. And often this is interpreted as you know, you can't be in pain, you're on your phone. How could you possibly be in pain? And this, I find across the board, regardless of you know, the age of the patient, where a patient is being doubted about whether they're in pain or not.

Emma: Subarna, I think that's so important that the cornerstone of sickle cell care is about recognising and treating painful crises. And what I find absolutely interesting is your idea that actually it is only parents and children and families who can understand what pain looks like, and that you as a doctor have to take their word for it.

So, you had a really interesting point that you were telling me that pain and the kind of pain that your patients experience changes over their age. Do you want to tell me a bit more about that?

Subarna: Yes, so I think it's got to do with where the bone marrow is active as you age. So, when a very young child experiences sickle pain, usually with bone marrow infarction, they may experience something called dactylitis, which is a kind of painful swelling of their fingers and toes. This is where the physical manifestation of pain happens in children. But as the child grows, the pain often, and this is not universal, there are some exceptions, but by and large, the pain tends to become more central, so the child will then start to experience pain in their limbs, and as the child grows into a teenager, pain often is more central. So, they may experience pain in their ribs or pain in their back, in their spines, or their pelvis, and so it's almost like it goes centripetal. I'm really generalising here. There are exceptions, of course. So, apart from dactylitis, where you can see the swelling, there is no other external manifestation of pain. So really, you have to believe the child or the family, who know the child really well when they say they are in pain.

Emma: Perfection. Alright, so if we're talking about painful crisis, your top tips are first of all, listen to children and their families, believe them. Give them pain relief within 30 minutes, go back and check if that's worked and if they need anything else. And I guess we also have to think about other things, so make sure they're not dehydrated. They've got fluids. Do they need IV fluids? It's November. Are they warm? Do they need oxygen? Do they need antibiotics? Is there anything else we need to think about?

Subarna: No, I think you've covered it all. I think it's important to focus not just on the pain, but everything else that's happening with the child. So, a child might be in pain, but it may quickly progress to an acute chest syndrome. So, it's very important to ensure that the oxygen saturations are monitored, and antibiotics given as appropriate, and suddenly, this time of the year, you're looking to explore whether the child has any viral infections and support as necessary. And yes, hydration is extremely important as well, and keeping the child warm. Yeah, all of that is very important.

Emma: Of course, I'm an infectious disease doctor, so I'm always interested in antimicrobial stewardship. Now, if we have good diagnostics, we know that a child with sickle cell crisis has got flu, for example, I'm still a little nervous not to give them antibiotics, at least oral antibiotics, but I've got no evidence for that. Do you have any feelings? Because I'd love to give out less antibiotics, but I'm always too nervous. I end up giving them oral antibiotics. What do you do?

Subarna: I must be honest with you, I do the same. I might send them home on oral antibiotics even if they are positive for flu on the nasal swab, partly because, I guess I'm telling myself, I want to stop them from getting secondary bacterial infections. I mean,

they are a very vulnerable group of patients, and we see pneumococcal deaths all the time, I have to admit.

Emma: Yes, I think this is a really good point. And I think this is an area where better diagnostics and better tests that would really reassure you and me there were no secondary bacterial infections, would make us a bit less nervous not to give them antibiotics at every point. And I guess in the future, what you would like is that your patients could actually test at home and decide if they had bacterial infections, if we had point of care blood tests that patients could use, but that's in the future. So, for now, pain relief, oxygen, warmth, antibiotics and just really, really good care and paying attention to detail.

Subarna: Absolutely, and may I add that actually universal vaccinations with Prevenar, and we also add Pneumovax, which is the PPV 23, has actually been shown to make a huge difference in outcomes, particularly with pneumococcal related deaths. So, part of good care, preventive care, vaccinations are very important.

Emma: Absolutely. And flu vaccine. Flu vaccine for everybody, perfect.

And so, I think let's ask a little bit slightly different question. You know your population really well, but most people don't see as many children with sickle cell as you do. So, what do families and young people want from a paediatrician? What can we offer them in terms of best quality care?

Subarna: I guess every paediatrician in the UK now will at least have one or two sickle patients in their clinic, just given how, widespread, people live everywhere in the country, so I think it's really important that, you know, paediatricians have the understanding of this condition. And also, I guess that relationship that develops between a child with a chronic illness and their family and the doctor and the specialist nurse or the multidisciplinary team that supports this chronic illness. So be it type one diabetes, be it any other chronic illness, and sickle cell is no different. And I think what families want is that kind of ongoing relationship, the relationship of trust, the relationship of kind of mutual understanding, learning from each other and supporting the family, and when done well, I think patients and families really value that.

Emma: I think that's really good advice. So even if you only have a few children that you look after with sickle cell, do your homework. Know a bit about it. And obviously we often say, just get in touch with somebody, somebody who sees a lot.

Secondly, any child with a chronic illness, it's about that relationship with the team, that MDT team relationship. So, listen to them, listen carefully, respond to what they want, and grow that relationship, because that's what matters to parents and families, that they

have a trusted person that they can always turn to. Really good information. I think it's really easy to forget it's a lifelong illness.

Subarna, I think this brings up a really interesting point. You're in London, and you look after a really big population of children and families with sickle cell, but up here in Newcastle on Tyne, we see very few, but there's a really good system of hub and spoke. So, do you want to talk a little bit about network?

Subarna: Yes, so for the last five years or so, there has been a real restructuring in England of the way sickle cell care is developed through managed clinical networks. And the whole of England has been divided into 10 networks for sickle cell and they are called Hemoglobinopathy Coordinating Centres (HCCs) and essentially there is a hub of kind of governance, guideline, provision and clinical MDTs that happen centrally in these networks, in these HCCs. And every hospital within a defined geographic area within that network will have direct access to those MDTs. So it is now very well established that if you are a single handed paediatrician in a very low prevalence area where there is barely any sickle patients, and you come across a really complex patient, or you come across any problem with it, there are very clear lines of contact that you can make and find advice very readily.

Emma: Fabulous, it's always best to ask the expert, and then having people like you so accessible really makes a huge difference. I think that's a really important point. Just ask for help. Don't do things by yourself.

That's brilliant. So, we've talked about how the networks operate, so let's just have a little chat about the actual management of sickle cell disease. So, we've talked a bit about the acute management of painful crises. But as we've discussed, this is a long-term condition. So, what kind of options are there for children and families with sickle cell?

Subarna: Yes, so although we've been working on new drugs for a long time, one drug that we've understood and learned about a lot is hydroxycarbamide, and it's been around in the sickle cell space for over 40 years. But in the last five years or so, we have had evidence that has really changed the way we do paediatric management of sickle cell disease using hydroxycarbamide. So, one thing that is now very well embedded in clinical practice, based on a lot of good research, is offering hydroxycarbamide to patients very young, nine months onwards, and even asymptomatic children with sickle cell anaemia. So, this is haemoglobin SS, and haemoglobin S because Cirrus thalassemia. So, if you've got a patient with SS or SB to zero thalassemia, and they're over the age of between nine months to 12 months, there has to be a very good reason why they should not be on hydroxycarbamide. So that should be the norm. Just like penicillin is the norm, hydroxycarbamide beyond the age of nine to 12 months really should be the norm. And these are the kind of conversations that should take place at the first consultation that you ever see with that patient, you know, the three-month-old baby that's come through

the screening program. You've seen them to start penicillin, you mention hydroxycarbonate.

Emma: That's so important, because I think this idea that prevention is always better than treatment, thinking about putting things in place before you hit crises and symptoms. So really, really interesting that is much younger than we've been used to. And just because this is my pet favourite topic, do you teach them all to swallow pills really early as well?

Subarna: It's a very good question. Yes, we have been doing that. We were very lucky now that we've got a syrup which is very palatable. Only five years ago, it used to smell like urine because it was compounded in the pharmacy, hydroxyurea and but now there is a palatable syrup, but it's very expensive. So, we do have pill schools. Our pharmacy where I work has a pill school, and it is really encouraging patients to start taking pills. And I think we've got a lot of uptake on that, I highly recommend that actually.

Emma: Yeah, as we promote KidzMed and pill swallowing a lot in Newcastle, and we actually routinely would suggest that children from the age of four, so anybody four and over swallow pills.

Subarna: Parents like that as well, particularly when they're traveling, it's much easier to carry pills than to carry bottles.

Emma: Cheaper, safer, easier to dose, less trips to go and get special medicines, because you can often only get the formulations from big centres. So, that's really, really interesting, that you are using these drugs at a lower age. What other new options for treatment can be offered for these families?

Subarna: One thing that has recently been approved in the NHS, and in fact, NHS was one of the first to approve this is gene therapy, and this is now available for children with sickle cell disease from the age of 12 and upwards. And this has been phenomenal, in the sense that for children who need cure from sickle cell disease but do not have a suitable donor, now have hope of cure.

Emma: So I think that's really important, that one of the options is a hematopoietic stem cell transplant, and that's actually been something that has happened for many years. How many of your children from your clinic get transplants now?

Subarna: Not that many, I have to say. So, we look after about 500 children at any given time with sickle cell disease, and annually, we probably send about three or four children for transplants, and these are children who generally tend to have very severe, chronic and progressive cerebrovascular disease. I think increasing use of hydroxycarbamide has made a big difference. What has happened is that they are not so ill during childhood, but we kind of have pushed it across to the adult age. And so quite a lot of the adult patients,

young adults, are now getting more symptomatic with chronic pain or acute pain not responsive to hydroxycarbamide, and they are often needing transplant when they become adults.

Emma: Interesting, so we've gone through these different treatment options, early hydroxycarbamide, stem cell transplant for the most serious cases, which is now happening more in young adults, and for those who don't have a donor, gene therapy, which potentially, in the future, could be really good, because potentially you might have less side effects, and that might be simpler to give

Subarna: Absolutely, right now, what happens is an ex-vivo procedure. So, you have to take the stem cells out of the patient, give the patient myeloablative chemotherapy, and return the gene edited stem cells back to the patient. But in the future, it might all be in vivo, and I can imagine a time when you might need the odd injection from time to time, and your sickle cell is killed.

Emma: That is incredible, so exciting. Really, really interesting conversations that you're going to have with your patients in the future. So, we've talked about acute crisis, we've talked about different managements, we've talked about what patients want. Is there anything else you'd like to touch on?

Subarna: Yes, there is one thing that I think trainees might benefit from going back and looking up, is the No One's Listening report, which I think it's been a landmark report, I must say, in the sickle cell sphere in in the UK. So, this was published by an all-party parliamentary group for sickle cell and Thalassemia about four years ago now, so November 2021 and it basically the topic is no one's listening, that's what the report says. And this exactly means that it came out of avoidable couple of avoidable deaths amongst young adults with sickle cell. And it is basically, not only just telling how the state of affairs is, which was terrible four years ago, but also some really helpful advice on how to improve services. And I think it's worth giving that report a read, because it gives you very good ideas on how you can improve, with very minimal financial input, how you can improve care for your patients. And that requires empathy, consideration, listening, learning, humility, all of that, and that has actually triggered quite a lot of improvement initiatives by the government. And that is very welcome.

Emma: Absolutely. It sounds like something that everybody should read, the No One's Listening report from November 2021 which is actually driven by patients, and a roadmap on how you can improve your own services. Listening to you, it feels like all diseases need to read this report, not just people with sickle cell!

Subarna, it's been such a pleasure to talk to you. I feel like I've done all my homework now you've told me all the things that I really needed to do to improve my knowledge, to think about how I could improve the services locally, and just to think about what's best for

patients and families. Before we go, we give everybody, it's a bit like Desert Island Discs! We give you the chance to give us three top tips. So, what would your three top tips be to all those paediatricians, clinicians, healthcare practitioners who are going to listen to this podcast be?

Subarna: So, one thing I would always say is, if you are lucky enough to train or work in a centre, or have access to a centre where there is a sickle cell clinic, go and sit in a clinic on your study education day, because seeing patients when they are in pain is very different from seeing patients when they are well. And you can then see the human side of them and that you will realise that these are just ordinary kids with aspirations like any other. And I think it's very important that you do that, and this is specifically for trainees. That is a top tip.

The second is, I guess, understanding, and this is more just to do with sickle cell, understanding that sickle cell is, to a certain extent, it's a hypostatic condition, so children are vulnerable to infections and so on. So, bearing that in mind.

And the third thing is that there is that inequity in care, without a doubt. You know, we know that there is lack of investment in research, there's lack of investment in care and having that awareness, and particularly with a group of patients who are already very societally sort of underprivileged. So, have that kind of understanding and empathy about the condition. It's very difficult to say people to change culture, but you know, just to understand that people can come from different circumstances, and they may not be similar to the ones that you're used to. Am I being preachy?

Emma: No, not at all. I love that. So, I think your three top tips are so important. I mean, go to clinic, absolutely. Go to clinic, see what children and families lives are like. Remember that children with sickle cell are a really vulnerable population. They're vulnerable to pain, they're vulnerable to infection, and most importantly, they're vulnerable because there is an inequity of care. There's a lack of investment. These are often children from marginalised groups. They're from different ethnic backgrounds. Maybe English isn't their first language. You need to be a sickle cell champion for these children. They need people to stand up and shout out and look after them, and I guess I'm just saying that's their job, isn't it?

Subarna: Absolutely.