

Airing Pain Programme 94: Complex Regional Pain Syndrome – Part 1

This programme was funded by grants from the RS MacDonald Charitable Trust and The Hospital Saturday Fund.

The first of two programmes on complex regional pain syndrome, or CRPS, which can be described as continuous and sometimes debilitating pain that can be confined to one limb, but has been known to spread to other parts of the body. CRPS is poorly understood, and no direct cause has been identified.

*In this edition of **Airing Pain**, Paul Evans speaks to Sunny Boshoff, author of CRPS Awareness: Moving Against Pain, who has had her own experiences with the syndrome, describing the agonising sensations she felt while living with CRPS.*

Professor Frank Birklein, head of the Peripheral Nerve Disorders and Pain Research and Treatment at the Department of Neurology, University Medical Centre Mainz in Germany is one of the world's leading authorities on CRPS. He explains the meaning behind the syndrome's name, what can cause it, and how understanding of the disease has progressed.

Clinical lead for the complex regional pain syndrome service at the Royal United Hospitals in Bath, Professor Candy McCabe, speaks about her experiences of treating people suffering from the disease, how it effects their emotional wellbeing, and the psychological therapies used to help people manage their pain.

Paul Evans: This is **Airing Pain** a programme brought to you by Pain Concern, the UK charity providing information and support for those of us living with pain and for healthcare professionals.

I'm Paul Evans, and this is the first in a series of programmes about complex regional pain syndrome, or CRPS. It's funded by a grant from the RS MacDonald charitable trust.

Sunny Boshoff: If you imagine toothache in a really bad abscess or something in your tooth and the sort of intensity of that kind of pain, if you can imagine that in your whole arm day and night, for the foreseeable future, that's the kind of pain that you'll get with CRPS. It ranges from being incredibly hot, to freezing cold, to feeling like somebody is pouring boiling water over your skin and then holding your burnt hand in a draught. So it's very complicated the kind of pains that you get with it. They're layers of pain as well, you will get deep pains in your bones, in your muscles, in your tendons, and then you'll have this pain on your skin. I kept going back to the hospital and saying my cast is too tight, I'm in agony my cast is too tight, because that's what it

felt like, and my hand was very swollen. But people kept looking at it and saying no it's fine, it's fine. And I was thinking it can't be fine, everybody else is sitting around here with casts on and they're not crying and I was in agony.

Evans: That's an extract from an earlier edition of *Airing Pain*, number 23 to be exact, in which Sunny Boshoff, author of *CRPS Awareness: Moving Against Pain*, talks about living with complex regional pain syndrome. It's a condition in which a person experiences persistent, severe and debilitating pain. Although most cases are triggered by an injury, the resulting pain is much more severe and longer lasting than that original pain.

The pain is usually confined to one limb, but it can sometime spread to other parts of the body. The skin of the affected body part can become so sensitive that just a slight touch, bump, or even change in temperature can provoke intense pain.

One of the world's leading authorities on CRPS is Professor Frank Birklein. He's head of peripheral nerve disorders and pain research and treatment at the Department of Neurology at the University Medical Centre in Mainz, Germany, and is the leading author of the German guidelines for CRPS diagnosis and treatment.

Frank Birklein: Complex regional pain disorder is just a denomination which was created because at the time when it was invented the pathology of complex regional pain syndrome was fairly unknown. In the meanwhile, we know a little bit more. So complex means that the symptomatology is complex, so the signs which can be seen in patients is complex. For instance, if you have a nerve legion then you have a pain, you have numbness in the nerve innervation territory but in CRPS a limb which is hot, other patients it's cold, sometimes it's sweaty, sometimes it's not, so it's a complex clinical sign. Nevertheless, it's regional because it's not confined to a nerve territory or a particular innervations territory, and it's not all over the body, it's confined to a limb. Sometimes it might be spreading to the other side. And pain is clear. And syndrome means it's not a clear cut disorder, it's a bouquet of symptoms and signs which characterise this syndrome so it's not a disease, it's a syndrome.

Evans: So complex, yes, the regional, where does it affect people?

Birklein: In the distal parts of the limbs, the feet or the hands. All other parts I would say it doesn't exist, probably there's one exception, the knee, but I would be very cautious to call pain in the hip complex regional pain syndrome, or in the ear, or the elbow, or the shoulder, or something like that.

Evans: Is it confined just to adults?

Birklein: It happens in children, it happens in adults, but the typical CRPS patient is a middle aged woman shortly after the menopause. This might have something to do with inflammation regulation, because women are more prone to develop inflammatory autoimmune diseases, and this is, in particular, true after the sexual hormones went down, because these are steroid hormones. You might have a trait, we still do not know what it is, but it could be that some patients are simply prone to get it, and we have to identify and acknowledge it early in order to help them.

Evans: Is that an issue as well, people can't identify it?

Birklein: Yes, people ignore it I would say. If there is a surgery, then surgeons sometimes got the impression that they have done something wrong, but they did not. So this is what has been told to the physicians, that nobody is guilty if there is a CRPS coming up. It's a trait, it can happen.

Evans: So, what causes it?

Birklein: This is the question that could fill the whole evening I would say, usually it starts with post traumatic inflammation, and this means that everyone of us gets an inflammation in a limb if he or she has a trauma. And usually this inflammation is shut down by our body within a certain timeframe, it depends on the kind of trauma you experience and so on. And in these patients the shutting down of the inflammation is delayed, or it is ineffective, so it is incomplete. And this means that you have a permanent inflammation going on in your limb, and this means that you have pain related to the inflammation, this means in particular these movements are related pain. And if you have a very fine orchestrated tool like a hand and you do not use it, and you have strong inflammation going on then you will have a loss of function. And as long as you lose your function the representation of your hand in your brain becomes smaller and smaller, as you know there is so much information that has been processed that comes from the hands and if this information is missing or is distorted by pain or something like that, then this has consequences. The other way round, so if you train a lot, you learn writing, if you have pain and you cannot move your hands or fingers intentionally anymore you lose your abilities.

Evans: I think you said the inflammation stays?

Birklein: Yes.

Evans: So is it something you see?

Birklein: Yeah you can see it, very often it's tremendous edema, and it's skin temperature difference and you can feel the sweating, and swelling also is a consequence of inflammation we know and have learned in the meanwhile. And the skin colour is different. So the definition of inflammation is swelling, it's warming, the colour, the red colour, and it's loss of function, and it's pain. These are the five cardinal symptoms of inflammation, exactly these symptoms occur in CRPS.

Evans: So is it that the original injury has gone, but somehow the repair process, if you like, is not turned off?

Birklein: Exactly, this is it in the initial phase, and after that all these consequences about losing the ability to use the hand, contractures, rearranging the brain, all these consequences take place the longer the pain and the inflammation persists. After a certain timeframe, I cannot tell you exactly when, but approximately I would say six months, after that the inflammation becomes less and less important but the brain takes over and causes the symptoms which are then present there for movement disorder, and the numbness, and the pain.

Evans: What's the process then for you to manage these people?

Birklein: To keep them going nevertheless, so I have to convince them that it is important to move the limb. I can give you an example, if you have children with CRPS it looks exactly the same, and you can treat them with drugs and so on, it will not work. But if you convince them to move the limbs, because there is something which is at the end of the tunnel, for instance sometimes I make negotiations with the parents that there is a pet waiting if they manage to use their right hand for writing, then you cannot imagine how intense these children work in order to use the limb more. And after half a year, or nine months the CRPS has gone away. I have some children which really go back to competitive sports and do not have any problems.

Evans: What's the outlook for most people who have CRPS?

Birklein: Ok I think I would say two third improve.

Evans: Improve, or...

Birklein: Improve. The least get pain free, only very rarely that people get pain free. You cannot answer this question very easily, because if I'm retired and I have some problems with my left hand and I try to improve it and reduce the pain, and then the patient says ok I can cope with it

because I do not have a problem to rest my left hand for half an hour if I have too much pain while using it. But if you are a hammersmith and your right hand is affected then it could be a very different situation I would say, then probably you lose your occupation and this makes a difference. So if I'm able and working in an office I can cope with the symptoms, I can cope with the pain then it's not uncommon that the patients go back to work. And they do not need any drugs or something like that in the long run. And most of them have some kind of loss of function, like contractures, but usually they are not so important. So they cannot deliver the power which they have been before, but I think this is in a lot of diseases, so if you have surgery on your gut or something like that, you have to acknowledge that you cannot eat anything anymore, and the same with CRPS. But this has something to do with coping and something to do with adjustment to the loss of function, and for some people it's easy and for other people it's simply not possible. And for those which it is not possible they suffer hard and others are ok I can accept that loss of function, but this will not affect my life.

Evans: So I would guess the treatment, as in most chronic pain conditions, the psychological treatment is important.

Birklein: Also important, yes absolutely, the pharmacological treatment, and we need psychological treatment

Evans: So as Professor Frank Birklein's experience backs up pain is not a fixed reality. This was the headline for a talk given by Professor Candy McCabe, she's the clinical lead for the complex regional pain syndrome service at the Royal United Hospitals in Bath and I'll be exploring that service in the next edition of *Airing Pain*.

Professor McCabe's research interests lie in the mechanisms and treatments for chronic pain.

Candy McCabe: All of our treatment guidelines, British Pain Society treatment guidelines for chronic pain, for pain management, is focused around helping people to live with pain, and helping them to function despite their pain. We of course have medications, we have spinal cord stimulators, we have fantastic pain management programmes, but we know that medications and spinal cord interventions and other neuro modulation will perhaps reduce the pain a little bit, take the edge off a bit, so that people are less distracted by it, less disabled by it, and can get on with their lives. We also know that chronic pain management programmes are excellent about helping people to live life with their pain. And often in pain management we talk about trying to walk with pain beside you rather than in front of you and encompassing you. Our

outcome measures for chronic pain programmes tend to be about have we improved function, have we improved quality of life, have we improved people's self-management skills, their ability to cope, their mood and their anxiety. All of which are really, really important, but when I talk to patients in clinic and say what are your goals for treatment the very first thing they say is I want you to get rid of the pain. They don't say I want my function better, I want you to reduce the pain a little bit, their top thing is 'I want this pain to go'. And at the moment in chronic pain we hedge that a little bit, we say we can't get rid of the pain but we're going to give you ways that you can cope with it.

Evans: So that's a barrier in itself when patients come to you, cure or self-manage, well obviously we want a cure, and the pain management techniques do reduce pain, but not cure.

McCabe: I think that's a fair comment, I think it would be a brave clinician who said come on our programme and we'll cure you of your chronic pain. That isn't the message we give, that isn't usually what we are usually able to achieve. So we're already having to not quite do what our patients really want us to do, and perhaps we can change that perception a little bit.

Evans: How?

McCabe: There's been a lot of publications in the non-pain field about how our bodies operate, and traditionally we used to think that our bodies sit there as receptive organs. That we wait for the environment to come to us and then we interact with that environment. And that we would learn through those interactions, so as a child you would learn how to walk and how to mimic things by interacting with that world. The assumption was always that it was very much that we're response organs, that we did whatever happened to us, but over the years that's changed and we now know that, actually, we see life, and we interact with life much more about how our bodies perceive that world around us rather than what may actually be there. So that's because we run on hypotheses, we run on assumptions about what the environment will be like. So if I can give you a simple example, if you were walking into a hotel lobby you would have a rough idea about what to expect there, because you'll have been, perhaps not exactly that hotel, but you'll have been in other hotel lobbies. And you'll know roughly what a reception desk should look like, and you'll head that way. So your behaviour in that new setting is very much influenced by your past experience. And you will also know on the very small level, that if you're going to walk on a carpet you would fully expect that that will be a softer sensation than if you were walking on hard floor. And so the muscles and the sensory networks, in order to interpret that information, will already be ready to accept that information, before you even do that action.

So if for example you walked on a carpet that you were sure was going to be a soft pile, then you would be ready for the relevant muscles for your feet to sink into that carpet, and also if you were there in bare feet your sensory system would be ready to feel that. But if by some clever trickery that carpet wasn't soft pile, actually it was hard, or perhaps even painful, the first reaction is surprise. It isn't what you expected, and therefore you have to change your expectations.

So what we think happens with pain, is that for those where the pain persists, you start to get set in a network. There's an expression in neurology that what fires together wires together. So if something has previously been working in that way, then it will continue to work in that way, because that's the quickest route. So if I gave you an analogy of a river, you start off and if there was just a little trickle of water going across the floor, that first trickle has to go where the floor takes it, where the natural dips and rises in that floor go. But if that water ran for the next 20 years, actually the floor would be carved out in the route where that water had been and the quickest way for the water to travel is down that channel. That means that all the other bit of floor in that room is no longer accessed, and there are wastelands of potential sources of information, or floor space in this scenario, which are still there, but we don't use.

Evans: If I have pain, the expectancy is that I will follow that path that I know that I have been before, does that make sense?

McCabe: Absolutely. So your brain will be wired to, if it's painful to move your arm, your brain will expect that no matter what you do with that arm, it will be painful. And because you know that you can't perhaps lift a cup or anything, then your brain will know that you will be weaker doing that, that it isn't possible, that you'll be clumsy. But also of course that all comes with emotional consequences. So associated with that pattern of behaviour is a level of perhaps anxiety, about 'will I drop something', or depression even, just frustration. So you get set in these pathways. But equally perhaps in our patients who have neuropathic pain, where if you touch the painful area it's intensely painful, and they can only feel pain, so they've stopped feeling light touch, pressure, vibration, they just feel pain, so what the analogy is, is that the information sitting on those river banks is that lost information, and in order to change the course of a river takes a huge amount of work. So what we are suggesting is that if we get people to think much more about where their limit in space, what the normal sensations will be if they were doing something, so let's go back to the example of you've got a really painful arm, what we want to do is to help people relearn what normal sensation feels like. So we would

work with them by asking them questions on the unaffected arm and say, how does this touch feel, how do you know it feels like that, where on your arm am I touching you, whereabouts is your elbow or your hand in relation to that. We learn by questioning, we learn by having to use our brains to consider information and come up with responses. Once you've relearned that skill, which isn't something you would naturally do, as children we probably explored that greatly, know about the different sensations, but as adults we have to relearn what those textures are and those sensations are. Then move to the affected side and say, ok before I touch you I want you to now be aware that I'm going to touch you, you should feel the same sensation you did before, try and find that information, try and access that information underneath the pain, because it's there, try and find it. And it's really extraordinary how people can suddenly find that information that they thought was lost to them, and lo and behold you start to shift the focus from pain into normal sensation.

Evans: The learned sensations, if you like, holding a cup of tea if you have a bad hand, well what I would do if I had a bad hand, if I found holding a cup of tea painful, I'd just use my other hand. Surely that's the sensible thing to do?

McCabe: Completely, because we're animals we have to keep ourselves safe, so all of our adaptations are about minimising risk to ourselves. The most sensible thing is to use that hand that works, but that then means that you have effectively lost a limb. So in our new therapies what we would say, ok so what we want to do is to hold that handle, now really be aware of how do you know you're holding the handle, what's the temperature of that handle, what's the shape of that handle. Rather than thinking what's my level of pain, so it's finding all that other sensory proprioceptive information that is normally lost to us, and masked by the pain. And by working in that manner, and as you can imagine it's a slow process, but patients absolutely understand it, and usually within a week of having really directed questions by the therapist they start to get it, and say actually now I'm going to look for those pleasant sensations, with those pleasant memories, rather than when I go to do something, looking for pain. So if pain starts to come back say actually no I want to go and find those nice sensations, those nice memories again. And you're effectively finding the information sitting on the river banks.

Evans: I still find it a very difficult concept to get hold of, how do you persuade somebody with chronic pain that they can rethink everything that they've done since they've had the chronic pain?

McCabe: In the first instance we'd get people to think back to a memory associated with that painful part, before the pain started. So that could be that if you've got chronic back pain actually remembering how lovely it felt having your back massaged with sun lotion while you were lying on a warm beach. So we want people to go back to that point. We know that chronic pain involves your emotional, your behavioural, your sensory systems, so we have to use all of those systems to change it. So if you were going to remember having a massage on the top of your back, first of all you'd like the therapist to put their hand on the non-painful side, now can you remember how that felt, tell me what the sensation was like, what were the sounds, the smells of that time, how many fingers am I touching you with now, can you find that information. Now I'm going to move my hand to the other side, I want you again, before I put my hand there, to think back to those lovely memories, remember how it felt, the sun, the seagulls, now I'm putting my hand on there tell me again, how many fingers have I got, how does that feel. And it's extraordinary how going back and finding that other associated information, you can start to unwrap all those other memories.

Evans: So does the pain go, or just the way you perceive it?

McCabe: Well, really excitingly, pain seems to go. Particularly our patients with complex regional pain syndrome where we really cannot touch their painful limb. But we've found that people can start to tolerate non painful touch while they are thinking about other sensations such as what direction is the sponge being laid in, how many fingers have I got. And they say, do you know I never felt pain at all. And there's a certain concern, 'did I make all this up', how is it that I've had pain for so long if something so simple can have made that pain free. And that seems to be more of a problem, you know pain isn't anything that anybody would wish upon themselves, or try and create. It's a miswiring and really all we are trying to do is get the brain back to a normal expectation.

Evans: Some of it sounds a little like visualisation. That certainly works, but it's temporary.

McCabe: Yes, and equally sometimes in some particular pain approaches, like greater exposure we would say you just need to do all those actions and just ignore the pain. And some people can do that, but they can't do it for very long. It's really difficult to ignore that. Visualisation can take you so far, but visualisation is quite a passive exercise, nobody's asking you particularly, so how high were the waves that day, or how gritty was the sand at that point. You're not really having to really draw on past experiences and answer questions, and we know that learning is really all about having to respond to questions, and having to assimilate

information and respond. It's called the neuro-cognitive approach, you need that level of inquiry within it, and have it multi-faceted really. So visualisation absolutely, direct touch, stimulus, so rehearsal of how that felt, real life experience of how that was. So it's a package of stuff. So I'm not saying ditch pain management at all, I'm saying add this in to all the great stuff we already have.

Evans: Professor Candy McCabe.

I'll just remind you that whilst we in Pain Concern believe the information and opinions on ***Airing Pain*** are accurate and sound based on the best judgements available, you should always consult your health professional on any matter relating to your health and wellbeing. He or she is the only person who knows you and your circumstances and therefore the appropriate action to take on your behalf.

Don't forget that you can download all editions and transcripts of ***Airing Pain*** from Pain Concern's website which is painconcern.org.uk. There's also information on how to order Pain Concern's magazine, Pain Matters.

Now, we at Pain Concern need your help. Like all charities Pain Concern rely on the generosity of individuals and funding bodies to keep us going. Don't worry, I'm not asking you to make a donation, although we would never turn it down on our 'Just Giving' page. But in order to carry on making these programmes, we really need to know that what we are doing is of benefit to people living with chronic pain, your family members and supporters, and I can't emphasise too much how important it is that we also get feedback from health care professionals on how these programmes help you help your patients. So do please go onto the Pain Concern website and click on the feedback button to take part in our short survey. Have your say, because without your views we won't know what we're doing well, and what needs improvement.

Now back to complex regional pain syndrome and the psychological therapy Dr Candy McCabe has been working on and enthusing about. And the big question for those that have CRPS, that therapy, where and when can I get it?

McCabe: Well, we're very much only in the pilot stages and we have been hugely influenced and informed by a group in Italy, who are based at Santorso, and they use something called the Perfetti technique, which was perfected by Carlos Perfetti. And he had written up this proposal that actually we just need to relearn this information. But alongside Carlos Perfetti many others have been working in a very similar area, and we ourselves have been using visual illusions to

trick the brain back to seeing normal. And this really just brings together lots of peoples work and this is by no means something that we ourselves have come up with, it's very much that we are drawing on other experience and other skills to try and apply it in a practical way to our patients with chronic pain.

So we're just starting out on this journey, it's still very experimental, but it intuitively feels the right thing to do.

Evans: Thank you very much indeed.

McCabe: It's really exciting, really exciting.

Contributors:

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