

Airing Pain Programme 23: Complex regional pain syndrome

Understanding regional pain syndrome, and treating phantom limbs with mirrors.

Paul Evans talks to Sunny Boshoff about her experience of complex regional pain syndrome and learns more about the causes of the condition from Dr Bill Macrae. We also hear from him and Dr Joan Hester about their work counselling and treating patients requiring limb amputations, including the use of the Visual Feedback Mirror to help those who have phantom sensations in their amputated limb.

Paul Evans: Hello and welcome to **Airing Pain**, the programme brought to you by Pain Concern, the UK charity that provides information and support for those of us living with pain. This edition is made possible by a grant from Astellas Pharma.

Joan Hester: Many patients have sometimes pleaded to have a limb amputated because of the pain. Sometimes it is not appropriate to do that and they find it very difficult to understand why the pain won't go.

Bill Macrae: It baffles me that the medical profession and, I must say, the public in general seem to ignore the subject. There seems to be some barrier to recognising that losing a limb can lead to pain.

Evans: We will return to that later in the programme. But first to a condition that we know very little about. It is difficult to diagnose and causes extreme pain and distress.

Sunny Boshoff: I am Sunny Boshoff and after a nasty accident, in which I had a crushed injury to my right hand, I developed a condition called 'complex regional pain syndrome', which is otherwise known as 'reflex sympathetic dystrophy' or 'RSD', or 'CRPS'. If you imagine toothache in the kind of... like 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 is the kind of pain you will get with CRPS. It ranges from being incredibly hot, to freezing cold, to feeling as though somebody is pouring boiling water over your skin and then holding it in a draft, holding your burned hand in a draft. So, it is very complicated, the kind of pains that you get with it. There are layers of pain as well. You will get deep pains in your bones, in your muscles, in your tendons and then you will have this pain on your skin.

Evans: Did you get a clear diagnosis of what it was?

Boshoff: No. I think I got the first diagnosis... it was about a month after. What happened, though, was I kept going back to the hospital and saying, 'My cast is too tight. I am in agony, my cast is too tight', because that is what it felt like and my hand was very swollen. But people kept looking at it and saying, 'No, it's fine, you know, it's fine.' And I was thinking: 'It can't be fine; everyone else is sitting around here with casts on and they are not crying', and I was in agony.

Evans: So, what did they do about it?

Boshoff: Well, I mean the thing you have got to do, is just keep your limb moving. I mean, really, that was the thing that they were doing. And I said to the occupational therapist, 'Why didn't you tell me?' She said, 'We were doing everything that we could do anyway and some people get really frightened if they have that diagnosis.' She just said, you know, 'It's going to hurt you, but you've got to keep on moving, you've got to keep... that is the only way you are going to overcome this.' I mean I still do it. That was in 2007. I still exercise my hand every day, with an exercise ball. I wake up in the middle of the night and I exercise it. I am constantly working on it.

Evans: Do you still have it?

Boshoff: I am not sure that it goes away. Because I have to tell you that, I had another accident about 15 months after I had got control of the CRPS and I broke both my wrists. I tripped over a small hooped fence in a park. My left hand appeared to be suffering the same sort of shooting pains in exactly the mirror spots to the other hand, you know the CRPS. But it was an injury to my wrist which was *really* strange that I should be having the same pains. And I went to see an osteopath, who I had been seeing for the CRPS, and he did a lot of cranial work with me that day, and by the time I had got back home, the swelling had gone down, the pains had stopped and they never returned.

Evans: Do you know what causes CRPS?

Boshoff: The experts will say they don't *really* know what causes it. It's as though your nerves have been scrambled. The neurological messages that are going to your brain are scrambled. The nerves are just sending off these signals and they are scrambled and so you are getting a signal which is saying, 'boiling hot', 'freezing cold', you know, all these mixed messages.

Evans: Sunny Boshoff's pain started from a crush injury, but some people develop persistent pain following an operation. It is not necessarily related to the original condition or even to any failure in the surgery.

Doctor Bill Macrae worked in the pain service at Ninewells Hospital, Dundee.

Macrae: In Scotland and the North of England in the 1990s we did a series of studies on the epidemiology of pain and we found to our great surprise that about 11 per cent of patients coming to pain clinics said that surgery was one of the causes of their pain, and another 11 per cent, they said that it was *the* cause of their pain. So 22 per cent of people were incriminating surgery as a cause of their pain.

Evans: That is quite worrying is it not, that people have surgery to get rid of an injury, or an illness and they come away with something possibly far worse?

Macrae: Yes, and particularly if, for example, you are a man who has an asymptomatic hernia – so you have a hernia, you have got a bulge – but it is not causing you any problems and you go and see your GP who says, ‘Oh we can get that fixed for you.’ You go and have a hernia repair operation and you end up with chronic pain and you did not have pain beforehand. And, you know, that hernia was not life-threatening. So you did not have to have that operation. The latest studies from probably the world's leading centre on this in Denmark would suggest somewhere between 5 and 10 per cent of men having hernia repairs have chronic pain afterwards.

Evans: So, what is going on there?

Macrae: Well, nothing has gone wrong. I think there is something that is extremely important to flag up. The original paper in which we showed these 22 per cent of people who had pain after surgery, what we also showed was that 18 per cent of people coming to pain clinics had pain out of injuries – all sorts of injuries – and this has been shown in subsequent studies as well. So, injury often causes long term pain. And if you look at this from an evolutionary point of view, then it makes sense if an animal is injured – say it breaks its leg – it makes sense for it to be painful afterwards, because then it is going to rest and the process of healing can take place. So, having pain after an injury is obviously a good thing, because otherwise if you keep walking on your broken leg it is never going to heal.

Evans: It has a purpose?

McRae: It has a purpose. But, the way it works – why your leg is sore the day after your injury – is not connected, say, to the broken bone; it is because the nerves to that area have wound up, they have turned up the volume and the whole area has become much, much more sensitive. And that state of heightened sensitivity normally lasts for a period of time related to the healing of the injury.

So, in some cases, not all, but in some cases, certainly after an injury or an operation, it may be that what you have, is the nervous system that winds up and becomes sensitised by the injury, but then it does not wind down again after the healing. So you end up with a healed injury if you like – broken bone or wound from the operation or whatever – but your nervous system to that area is still wound up, so the slightest thing is going to cause you pain. If you think of it like a volume control and in fact we know that there are volume controls in the nervous system – both at the periphery, if you like, at the skin of the surface of the bone, but also in your spinal cord where the nerve has its first relay going up to the brain. There are volume controls throughout the nervous system and if these get turned up and then they don't turn down again, you can see how that might cause chronic pain.

Evans: So, how do you turn them down?

Macrae: Well, there are drug therapies that will help and the drugs that we commonly use for neuropathic pain – the tricyclic antidepressants like amitriptyline and some of the anticonvulsants like gabapentin, Pregabalin, sodium valproate – these drugs can help. But it is not always possible to turn the pain down. Any sort of neuropathic pain is difficult to treat and so chronic pain or surgery is really no different from those, these are difficult pains to treat.

Evans: Some of drugs you mentioned there, they are antiepileptic drugs?

Macrae: Yes.

Evans: So, what does that have to do with pain?

Macrae: Well, if you think of epilepsy as the brain being hyper excitable and so it fires off and causes a fit, and what I am saying is your spinal cord or your periphery might be over excitable and what these drugs do is they damp down over excitable nerves. So, what you end up with is the whole system being damped down and less likely to fire, less likely to cause pain. And the tricyclics are not working through an antidepressant effect – in fact they were originally used as antidepressants but in far higher doses – but at much lower doses they have an effect on some of the transmitters in the pain pathway as well, at the spinal cord level. So, they are not working because they are antidepressants, they are working because they affect nerve transmitters that are present in the pain pathway in the spinal cord.

Evans: Dr Bill Macrae.

One category of chronic pain after surgery is known as phantom limb pain, this can follow the amputation of a limb. Now there are various reasons why an amputation might be necessary. Some conditions like diabetes reduce the blood flow to the extremities and that can cause damage and I have known lifelong smokers – my mother included – who have had toes and legs amputated for similar reasons. Another reason is to prevent a tumour invading the surrounding tissue, but the major cause for amputation is due to crush injuries, and that is particularly relevant today with more and more soldiers returning from Afghanistan with blast injuries from IEDs (Improvised Explosive Devices).

Dr Joan Hester is a consultant in pain medicine at Kings College Hospital in London. She is past President of the British Pain Society and has also given talks and written articles for the Limbless Association, that's an association for amputees.

Hester: Phantom limb pain is defined as a pain that follows amputation of either a leg or an arm. You could also call it 'phantom limb sensation', which is the sensation that the limb is still there even though it has been removed. It is not always painful, but sometimes it can be *extremely* painful, usually if it has been painful before it was amputated, it will be more painful afterwards.

And it is because the limb is represented as a map on your brain – all your body is represented as a map and you have bigger representation for some parts of the body than others. So your thumb for instance, will have a big area, because it does so much in normal life, and your big toe will have a big area. So, when the limb has been amputated, the map is still on the brain and the brain slowly reorganises it, so that the limb gradually feels as though it is shrinking in your mind, until only the big toe for the leg, or the thumb for the arm feels as though it is sticking out of your stump. It is a little bit of a difficult concept.

But, that might take six months, that might take longer sometimes. And during that process some people feel the pain that they had before in the limb, although the limb is not there. And that could be very strange couldn't it, if you were not expecting it? So we try to tell people before the amputation that this is something that might happen.

And they do get other kinds of pain as well, because the stump itself... obviously when they do the operation, they have to cut through the bone, they have to cut through muscles and nerves and a good surgeon will bury the nerve under the muscle, because the nerve tries to regrow – when they cut into the nerve it sends out little feelers like tendrils of a plant and they can become very sensitive: what we call a 'neuroma'. So, you can imagine, if that was under the skin, especially where you were trying to wear your prosthesis, it could be *extremely* unpleasant and uncomfortable.

The third kind of pain is a general sensitivity of the stump, which often happens immediately after the surgery, where it is just very sensitive to touch, or to hot, or to cold and that is a kind of nerve pain as well. That can be very unpleasant and stop people from being able to wear their prosthesis. But prostheses have improved so much – they used to be very heavy, very cumbersome, lots of straps, and now they are very light and wonderful modern materials and they have suction legs or arms so that they just sort of glide on and stay on much more easily. So, I think the prosthesis technology has improved, but maybe our management of pain hasn't kept quite up with that.

To try and help the pain, of people who have had amputation, firstly, is to prepare them for the kinds of pain that they might experience, because I think understanding it does *really* help people. And it was something that was fashionable a little while ago, which was trying to reduce the pain before surgery, by giving them an epidural – this is particularly for the lower limb – in order to numb the nerves, so that even before the operation the pain was lessened and continuing that through the operation and afterwards. And there have been some different results. People have studied it, and one group of patients – ‘yes, it did help’ – and another group of patients – ‘it helped at the time, but not afterwards.’ And then we can give people the drugs that help what we call ‘neuropathic pain’, nerve pain – like gabapentin or pregabalin or amitriptyline – can start those *really* early, just immediately after the operation.

You can talk to the surgeons. Many surgeons are very, very good, because the way they do their surgery is fairly critical to the amount of pain that somebody will get. And we can obviously after the surgery, give very strong painkillers, like morphine combined with – if it is possible – an epidural to actually numb the area and that seems to help.

Macrae: When I first started back in the 1980s, 1970s, we still got people who had been told that they couldn't have phantom pain, particularly in the military. The military doctors seemed to be in denial about phantom pain and they regarded it as a psychological aberration. I've been looking back through history about this, and if you go right back to the 15th century, there are a lot of articles about, or paintings or things about, amputation; you can actually see in churches, paintings of saints, replacing amputated limbs. So, clearly people had amputations in those days. And then there was a French surgeon called Ambroise Paré, in the sixteenth century – he was the first one to talk about the phantom phenomenon, but he didn't call it that. So, it was talked about then and then, of course, Lord Nelson lost his arm and he had this classic feeling, that many amputees get, of his fingernails digging into his palm, which is often what people describe to me. Yet, you would think with people like that, it wouldn't be ignored; people would say, ‘oh yes, this is a phenomenon’, and yet it was still denied.

The man who coined the term 'phantom pain' was a wonderful man called Silas Weir Mitchell, who was a doctor in the American Civil War, although I am proud to say of Scottish origin, and he coined the phrase phantom pain and followed people through in the American Civil War – very interesting man, he was an neurologist – and he documented the whole thing, but still it wasn't recognised.

If you look at the First World War and you see how many people were killed, or injured in the First World War – there were 12 million soldiers killed – and huge numbers of people suffered amputations and yet the subject was again largely brushed under the carpet. We come to the Second World War and they are putting in incidence of 2 to 4 per cent of people, which we now know is far, far too low and decent studies – again, an excellent group in Denmark – showed at one time or another about 80 per cent of people who have had an amputation will have pain. They are not having it all the time, some of them may have had it for a short time, and it may not have been very serious, but a figure of 2 per cent is clearly completely ignoring the problem.

When you look at their papers, they are attributing it to psychological causes, or they are just not believing their patients. So, it baffles me that the medical profession and, I must say, the public in general seem to ignore the subject. There seems to be some barrier to recognising that losing a limb can lead to pain – I mean, 'if you haven't got a leg, how can it be sore?' as most people would ask. So, you can see if we can't recognise something that is as obvious as that, you know pain after a hernia operation, people are not going to be able to understand that. I think that the surgeons suffer from guilt because they think that they might have done something wrong, which isn't the case. I mean, I don't think you need to do anything wrong, I think, as I was saying right at the beginning, some people are going to have pain after an injury. It is bad luck, but it happens.

Evans: But it is a very difficult concept, isn't it? That, if my arm were removed above the elbow, to imagine that two foot away, there is two foot of air space, and I hurt at the end of that nothingness.

Macrae: Yes, it is difficult, but it happens. And, in fact, there is a lot of really interesting research, particularly brain imaging which has taught us a bit about it. It hasn't given us the answers we would like, but it certainly helped us to understand it better. And it may be that part of the problem is that, the brain re-maps and part of the brain still thinks that you've got your hand – or your arm and your hand – and other bits don't re-map and you have what the psychologists call 'dissonance' between the two bits of your brain, so that one bit is saying, 'you do have a hand', and your visual system is very clearly saying that 'you don't have a hand', because it can see you don't. Of course, dissonance can cause very unpleasant

sensations like sea sickness – that's dissonance between your visual system and your balance system. I mean that isn't the whole story – there is much more to it than that.

Evans: Can that incorrect mapping of the brain be re-mapped?

Macrae: There is some interesting work from Herta Flor in Germany about re-mapping and one of the things that she showed was that, if you give somebody an artificial limb that moves and a functional limb – you know they have these limbs with electrical motors and things in them – and they can look at it and see what looks like a hand moving where they think their hand is. These people tend to get less re-mapping. It's not cut and dried – it's not as simple as that – but there is a tendency for an early use of prosthesis – and particularly an active prosthesis like that – reducing re-mapping and reducing pain after amputation.

Hester: The psychological process is also very important and it depends how somebody's coping with their illness beforehand, or, if it is a sudden accident, whether they witness that accident or whether they have memory or no memory and how it is going to affect their lives, as you can imagine. I mean it is extraordinary with the soldiers as an example, because their motivation is to get back into active service again, but somebody who has had many years of dealing with poor circulation in their legs might not have walked for a very long time so that they become very depressed – their social life has gone, they have lost jobs, they have lost friends and they will become very depressed.

An amputation may or may not help that; it depends how... if they can get active again – that's the aim, to get active again. I think understanding pain, is incredibly important, pain patients have sometimes pleaded to have a limb amputated because of the pain. Sometimes it is not appropriate to do that and they find it very difficult to understand why the pain won't go, so we have to explain again and again sometimes that the nerves are still there between the stump, the spinal cord and the brain, so you are not cutting out the nerve pathway that tells the brain that there is something there and that's why the pain persists.

In trying to prepare patients for this after surgery, we can't numb the brain so much. We can stop the whole area becoming so sensitive with some of the drugs. It is also critical the way the dressings are applied and the way the stump is looked after, to prevent infection in particular. We can teach people how to use TENS machines, acupuncture has been used, physiotherapy is very important getting people walking as quickly as possible, and something has been used called 'mirror therapy'.

Now, this is putting a mirror on the bed or on a hard surface – a table if it would be the arm – opposite the good limb, so that the brain sees the good limb as the one that has been

amputated. And then the brain will work on that area and a physiotherapist can guide the person to do things with the good limb that helps the brain to translate that into good things for the limb that is not there anymore. It sounds a bit strange but it is helping the brain to re-map itself in a helpful way, but it requires a lot of patience and in busy hospital environments it is not often done – you would need a very helpful, friendly physiotherapist. Or, if you were taught how to do it yourself, you can do it at home. You can get yourself something called a mirror box, or if you have got a very kind person in your house they could make one for you! It is all quite simple and feasible, it's just somebody teaching you what to do and the importance of it.

Evans: Dr Joan Hester, Consultant in Pain Medicine at King's College Hospital in London. Now, as always, I would like to remind you that whilst we believe the information and opinions on **Airing Pain** are accurate and sound based on the best judgments 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.

Now back to those mirrors and to Sunny Boshoff, who, as you will remember, has complex regional pain syndrome.

Boshoff: During a course of occupational therapy I was kind of looking around the room while I was having my hand tweaked and I saw this kind of box thing, with a mirror stuck on it and I said, 'ooh, what is that?' And they said: 'That is a visual feedback tool.' And they said, 'would you like to try it?' And I said: 'yes, I would.' So they made an appointment for me to see somebody who had been trained in visual feedback and it is fantastic. What it is really, is a mirror, so if you imagine a vertical mirror, and you put your injured hand behind the mirror, and you put your uninjured hand in front of the mirror and you look into the mirror, you line it up so that when you look into the mirror you see two healthy hands. Then you try slowly to do movements with both hands and your eye sends a message to your brain, that you have indeed got two healthy hands and against all odds it helps the pain. I cannot do certain things – I cannot make a fist with my injured hand still, even with all the exercises and whatever – but if I do the mirror work and I make a fist with both hands, it feels as though I am making a fist with both hands, if I am looking in the mirror.

Evans: But you are not...

Boshoff: No.

Evans: But I guess with CRPS you need to do this fairly often?

Boshoff: Yes.

Evans: So how do you get around that?

Boshoff: A friend of mine made one for me, which was sort of like a table, in a sense, or a box, like a tunnel, and a mirror on the one side and you put your hand into the tunnel – the injured hand – and you would look at the reflection. It just became a big cumbersome thing, I couldn't carry it around. So, I designed one which is based on like those clipboards that you get – the sort of plastic clipboards that everybody uses. It has got a cover, so that you fold it around. So I manufactured one using that kind of material and the mirrors are on the inside and you fold it open.

Evans: So, my right hand is injured, I put my right hand behind the mirror...

Boshoff: That's right...

Evans: And by moving my good hand, in the face of the mirror, my brain is telling me that from the reflection of my good hand, that my bad hand is working well?

Boshoff: Yes.

Evans: For those of us who are a certain age, we will remember Harry Worth the comedian, Harry Worth where he went in front of a shop window and raised his left leg and then the mirror image looked as if he was raising both legs at the same time...

Boshoff: [Laughs]

Evans: And it is very similar to that isn't it? I am looking at my image of my good left hand, I am looking at the reflection of that and my brain is telling me that that is my good hand. So, maybe what it is doing is unscrambling those circuits.

Boshoff: It is very difficult when you are looking at the very injured hand – and it's purple and it's swollen, it's hairy and this sort of stuff – to imagine that it is okay. But behind the mirror when you are looking at the good hand, your brain does it for you, you don't even have to think about it.

Evans: Sunny Boshoff.

Of course the visual feedback mirror won't be for everyone, but it is suggested that the earlier you start using it, the more effective it could be. Sunny has also written a book about her experience of managing complex regional pain syndrome, it is called *CRPS Awareness*:

Moving against Pain. You can find out more information on it and the visual feedback mirror at her website and that is at crps-selfhelp.com.

Now, before we end this edition of ***Airing Pain***, let me just remind you that you can put a question to our panel of experts or make a comment about the programme on our blog, message board, Facebook, twitter, or via email and of course pen and paper. All the contact details you need and a link to download all previous additions to ***Airing Pain*** are at our website and that is at paiconcern.org.uk.

The last word to Sunny Boshoff:

Boshoff: After the accident my little finger, my ring finger and the middle finger were stuck together, I couldn't even get a towel in between them to dry myself, and they were very, very painful. It was quite hard to touch them or do anything, but I worked at it and kept on trying to get them apart and then eventually, I remember the day I went to the occupational therapist and I took some moisturiser and I said, 'Oh, I'm going to show you something.' And I rubbed the moisturiser in and I was able to put my fingers through each other with both hands. And it was so exciting, and you know they celebrated with me and I really encourage people to get involved with the people who are trying to help them because it is really a good way to do it.

Contributors

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