

## **Radio Phone Interview w/Maureen Coleman & Dr. Walter Kocha**

Interviewer = "Int"

Dr. Kocha = "K"

Maureen Coleman = "M"

Int: Well they're not only difficult to detect, but Neuroendocrine Tumours are the most rapidly increasing form of cancer in the world. Now that has the medical community acting to raise awareness about these so called NET cancers so that lives can be saved. To learn more about this, we are joined this morning by Dr. Walter Kocha, a medical Oncologist at London's Health Sciences Centre, and by Maureen Coleman, President of the Carcinoid Neuroendocrine Tumour Society of Canada.

Int: Good morning to you both

K&M: Good morning

Int: Good to have you with us. Doctor, explain in laymen's terms if you can, what these so called NET cancers are

K: Well the neuroendocrine tumours come from a particular group of cells called, not surprisingly, neuroendocrine cells and we actually see them as their own organ system now, although if you were to look at a body, you would for the most part, not be able to see these as a separate organ somewhere in the body. They actually either consist of an extra single cell or multi cells scattered throughout many parts of the body and they have a very specific function that usually includes a signalling function, it does signal things to occur in the gastrointestinal tract, the neuroendocrine cells in the stomach, receive signals and produce hormones to cause the stomach to secrete acid, and perhaps one of the more notable ones, the neuroendocrine cells that are scattered throughout the pancreas, the pancreas has a main function of producing digestive juices, but within that there's these islands of neuroendocrine cells that make such things as insulin and other hormones, so they are scattered throughout the body and they can go malignant or become cancerous; however, their characteristics are very different from most other cancers because a large portion of them when they do, become very slow growing tumours and take on other properties which allow us to take advantage of some of these properties in terms of treatment and because they have different properties, we have found that we have to treat them differently from most other cancers. Some of the rules of thumb or approaches that we use for measuring other cancers, can be different or modified because of the difference in these properties; they are of very slow growing character.

Int: So while they're slow growing they can be a bit of a chameleon?

K: Well that is certainly a problem that they very often don't let you know they're there with the exception of those that take on the ability to over-produce some of the signalling hormones that they do, the most common one in the gastrointestinal tract which have gone under the name of Carcinoid Tumour, an interesting name which actually meant cancer-like, we're now trying to get rid of that tumour, but if they ever produce some hormones that the normal cells would produce, people end up with considerable problems such as flushing and diarrhea, in fact the diarrhea can be so severe that the patients can't leave home, they always have to be near a toilet. And as well, if it happens to produce insulin, patients have terrific problems with low blood sugars. Not all of them do, some of them will and if they do, they may be found earlier; however, it is as well a problem that when they over-produce their hormone, they only usually are detectable because they have spread to the liver and are producing it from the islands of tumours that have

spread to the liver and are releasing it directly into the circulation. So it's a real problem because the darn thing often doesn't let you know it's there until it's taken on this ability to spread and has done so.

Int: And so how do you go about though detecting these? I mean, would a person who presents with, say gastrointestinal symptoms, would doctors immediately think well, maybe this is what we're dealing with?

K: No they wouldn't and there's a particular reason for this and in fact it has a lot to do with what symbol the patient groups utilize, the zebra, and that is in medical school we have and we continue to teach that when you diagnose things, you should think about the most common things. The metaphor is, when you hear the sound of hoof beats you think of horses, you don't think of zebras. Well this is one of those zebras and it's not one that people do think of first because it is so uncommon. It was uncommon enough that a cancer specialist who was seeing all sorts of cancer from a community, would only see two or three new ones per year, and probably most family docs would never run into one in their whole professional career; it was sort of that uncommon. Now they are increasing and that's less likely to be true but in this situation in which these symptoms which are identical to Inflammatory Bowel Disease or Irritable Bowel Syndrome, which is very common in society, might be thought of for these patients who don't respond well to therapy or who continue to have problems with. Otherwise they are sometimes found by accident and the most common places that a person has an appendectomy and there is a very small neuroendocrine tumour there, which is very often cured because of that and lastly, they can grow inside the small bowel and sometimes cause partial blockage or obstruction and therefore may come to surgery and be removed before they have taken on this ability to be able to spread.

Int: So does this increase the importance of having for example, a colonoscopy examination on a regular basis?

K: Well the answer is probably yes because of the gastrointestinal neuroendocrine tumours, the one that is increasing the fastest, is the one that is in the lower end of the colon and these will be picked up. And in fact, some people wonder whether the fact that we're seeing more of these is because colonoscopies are being done more frequently or picking up more of these, but yes, this is one of the ones that can be picked up by that particular procedure and it can occur in the stomach, and if people have problems in the stomach it can certainly be picked up by having the scope of the stomach. But there is no good scope for finding it in the small bowel and that is the most frequent place that it starts.

Int: So that's another problem.

K: Yes

Int: And can it also erupt in other areas of the body?

K: Yes it can, there are other varieties, the gastrointestinal tract is the most common one but for instance the centre part of the adrenal gland which makes adrenaline, those are neuroendocrine cells, which where the outside, by the way of the outside of the adrenaline gland what's called the cortex, produces steroids and is not a neuroendocrine cell. So you can get particular types of tumours from there and its gone under the name of Pheochromocytoma and the outshot of that is that they can produce adrenaline and can cause extreme problems with high blood pressure, they can occur in the lung and that is also rapidly growing for reasons we don't understand and there is a variety of a particular cell within the thyroid gland that will have that, so the abnormal tissues are widely dispersed over the body and in any cases, they can become malignant or cancerous. It has been a problem and within the medical profession and elsewhere that they are often thought of as benign tumours and the other term that is used is indolent tumours which are not very aggressive and that is often based on the fact that they don't grow very quickly. But in the end they are

cancerous and they can spread and a lot people eventually will die of them. Although, you can live with them for many years and sometimes decades, especially with our modern treatments that we have for them.

Int: A real challenge! Maureen, I want to turn to you. Maureen Coleman is President of CNETS Canada. Maureen, have you had any direct experience with neuroendocrine cancers – the cancers we’re talking about?

M: Yes I’m a patient

Int: You’re a patient?

M: I’m a long term patient

Int: Can you talk about how you learned that you had this. What was the course of this?

M: Well, it was with great difficulty that I discovered it. It took many years to discover that this is what I have, and I had one of the symptoms that Dr. Kocha has described which is flushing. I think I was probably a champion flusher.

Int: Congratulations Maureen.....oh dear

M: I was going from here to there saying can’t understand it, until eventually my G.P. said, have you ever had an abdominal ultrasound. This was in the year 2000 and I said no. So I had it, an abdominal ultrasound and what it discovered was that the ultrasound displayed that I had tumours in my liver, at which point I started seeing an Oncologist and I had a biopsy and I started going on the medication that many of the carcinoid variety patients take.

Int: So did you get a complete explanation though at that point then, of the particular kind of tumour were looking at?

M: A partial explanation because I was told initially that it was cancer-like but that was further confused because I was sent to an Oncologist. So I don’t think I thought about it really, I mean I just accepted that I’m seeing an Oncologist that must mean something. I think It takes a couple of years for a cancer diagnosis to be internalized anyway, so this was all new and I went through it but I slowly grew into that position of being a cancer patient. So what I heard about what I have was not exactly what I ultimately realized I had.

Int: So you came away with the impression that it wasn’t cancer, but maybe, it was sort of you were caught in the middle?

M: I was, yes. But later on when I consulted more physicians, and joined a support network, an online support network, and eventually we started a support group in Toronto in 2001, that was when I started to learn more and more from other patients and other sources of information.

Int: Were their stories basically similar to yours then, were you hearing the same stories told back to you?

M: Yes, the story of it taking years to become diagnosed, that was extremely common. And also being told it was in your head. So people would be telling others that they were told that they were imagining what they had. I mean, I know for myself that I saw psychiatrists trying to figure out what was going on.

Int: You were that desperate?

M: Well I was curious. I couldn't understand the flushing.

Int: Was that the only major symptom you had?

M: Yes, for me that was my main symptom. I didn't really have many of the gut issues that some other patients have. I was flushing from head to foot.

Int: So let's talk about your organization because, when was CNETS Canada born then?

M: The support network was born in 2001 but CNETS Canada became a charity; well it didn't really become a proper charity until 2008. It became incorporated in 2007 and we had a Board of Directors and we had members which consisted of at the time, of support groups. So we had support groups all across Canada with members who vote for the Board of Directors, who are involved in the network in one way or another. And this last Cancer Day, Worldwide NET Cancer Day, some of the people who are support group leaders and some others, went to hospitals and set up an information table; one did it in BC, another one did it in Alberta, there's gonna be one in Ottawa still to come, and also in Toronto and in London. I'm going to be in London tomorrow with a table.

Int: Dr. Kocha, this organization must be a great benefit to you and other Oncologists who are trying to get the message out and trying to explain what's going on here to patients?

K: Yeah, they certainly are, they hold sometimes very large meetings in a region where we get to talk about it, so it is of importance, but mainly it's important to the patients. It is a problem that most of the rest of the population doesn't know much about it, and I think there's a much more generic problem that occurs with these especially in terms of the treatments that we can offer in this situation or in fact other situations. I think we have a real problem in Canada dealing with various diseases that are low in incidence and we often call these orphan diseases. So although the common tumours, for instance, get a lot of attention paid to them, and a lot of trials are done to them, very much less occurs for the low incident diseases. Companies in general do not see this as a very profitable area to get into because they are so limited in numbers, they don't see a big profit capability for all of these diseases and I think it speaks to that this disease and a whole number of other low incidence orphan diseases, that we need a strategy in Canada for dealing with orphan diseases. Such a strategy occurs in the United States and other countries, but we have never formulated one for Canada. And it needs it so that these group of people can get their due and can get the sort of therapies available which are sometimes very useful to these but are otherwise very difficult to get because companies don't much care and the governments don't like to spend the money.

Int: We hear that over and over again, it's very sad isn't it?

M: Can I just offer one thing?

Int: Please do Maureen

M: We do not have a registry, we do not have statistics for this cancer, so we can't count how many people we might have. We're not classified under our own name Neuroendocrine; so we're invisible. This is one of the reasons that we're underserved, what we really, really need is to have our own name under the list of cancers and statistics collected on our cancer. I think that if that were to happen, some work could begin to be done.

Int: Absolutely

- K: I think I have to modify that a little bit because we can actually ferret out the number of tumours that occur but the large cancer organizations, Canadian Cancer Society, this sort of thing, when they count cancers, they count it on the basis of the organs in which they occur in. So very often this would be counted under gastrointestinal cancers; however, they diverge so much in behaviour that they are really different from other gastrointestinal cancers and Maureen is right, it does need to be reported separately from the other gastrointestinal or other tumours and to date it has not been. Although we can go back in to the registry's that occur and actually find the status, we have in fact been able to do that in Ontario and I've found since 1964 when it first started being collected, until about 2002, which is one of the last dates in which we have verified data, that the incidence (that is the number of new cancers) of these Neuroendocrine tumour groups, has increased from about 2 per million to about 22 per million in early 2003. We think it's now somewhere between 30 and 40 per million in terms of the rate of increase that was going on.
- Int: It occurs to me too Dr. Kocha and Maureen Coleman, that if we tend to give less credence and pay less attention to any form, any one form of cancer, aren't we perhaps down the road robbing ourselves of a better understanding of cancer. Who knows what investigation is going to lead to a major breakthrough in combating this disease on a large scale.
- M: You're absolutely right. That question is really appropriate.
- K: There is now somewhat more interest being paid to it and in fact, some of the recent strategies in newer cancer treatments coming along is based on the knowledge of what sort of differences there are in the cancer cell. Two years ago in the neuroendocrine tumours worldwide, there were absolutely no clinical trials available, to say that one treatment was better than another, by that I mean high level clinical trials which would we call perfectly randomized trial. As of today, we now have 5 clinical trials, perfectly randomized, and they are all positive, they all show benefits, some of the newer treatments that we have available and in some cases they are of a very considerable benefit. So that has changed to some extent, but yes that is a particular problem that not much attention has been paid to them. It is because we have knowledge of how they work and these trials that I say in order to get enough patients to populate these trials, they have actually been funded by the larger drug companies and they would have to be worldwide trials in order for us to get an answer within 2 or 3 years.
- Int: Maureen, you were going to say something?
- M: Yeah I also feel that the numbers could be way higher even than you suggest Dr.Kocha because where I live in Toronto, I live in a 10 min walk of 6 other patients.
- Int: Within 10 minutes of 6 other patients?
- M: Neuroendocrine patients. There's 2 to the East.....
- K: I think this is probably indicative of the other feature of this disease because you can count the incidences (that's the number of new cases), but the other thing that they determine and this has to be done mathematically from registry, is the number of patients who have the disease currently that are alive and do have the disease. So interestingly enough when they do these calculations on the tumour registry, it doesn't seem to matter where in the world they do this, that for gastrointestinal type of cancers, the Neuroendocrine Tumours are number two in terms of prevalence after Colorectal but they are higher in prevalence than stomach cancer, liver cancers, oesophageal cancers, and any of the other gastrointestinal cancers, they are higher in prevalence and that is probably a property of the fact that they are longer lived. Usually the incidence (number of new cancers) and the prevalence are very similar because the cancers can progress very quickly, and then therefore the numbers are very much the same, but in fact, the prevalence of

Neuroendocrine tumours is very much higher than a lot of other tumours simply because the tumour grows more slowly and a lot more patients are alive with it. It is a real problem for these patients by the way that they sometimes get regarded as cranky patients because what are they belly aching about there's are other tumours that will take patients away much more quickly and you have the tumour which is often called benign, this sort of thing, and you're doing a whole lot better, so why are you bothering us anyway. Patients do not like this and I think in fact that that's not ethical because in the end it is a malignant cancer and there are treatments that can prolong patients' lives with survival from years to decades and patients deserve a shot at that, they should not be ignored and told their lucky they don't have one of the other cancers.

M: I agree as a patient, and I'd like to say that London Health Sciences Centre in London, Ontario has developed a treatment plan that gives continual care for this class of cancers for this patient community and it means that they can have many options as they go along, as they progress through the disease. Or even as they can get '*inaudible*' with the disease but they have a number of options because of the team in London, Ontario.

K: I think that this exhibits another principle in dealing with these so called orphan group of diseases because they occur so infrequently that most practitioners out there will only see them occasionally and not get very much experience and these patients are far better served by being seen in a centre of expertise that see's a lot of them. And London I think is one such centre for Neuroendocrine Tumours in Canada and I may be blowing my horn, but I think we are probably the best centre in Canada for having a comprehensive multi-disciplinary program in this disease. But it's not the sort of thing included in that is other orphan diseases, such as we've seen casually by people who will only be seen once in a while. It should be seen by groups of people that are dedicated with that and see a lot of them, typically 100 -150 new ones per year and see at least 200 follow-ups per year. With that you get enough experience to really know how to deal with the diseases. But if you deal with less than that, you don't have the knowledge and especially the experience in dealing with and experience has turned out to be a very important part because then you develop the skills to know what's happening with the disease and then knowing what to do with them. The metaphor that I tend to utilize is that if you're going to get on an airplane, and you found out that the pilot had never actually flown the plane before, but he read the guidelines last night so it should be okay, you probably wouldn't get on that airplane and I think the same metaphor applies to the treatment of a lot of disease. If they are low incidence diseases, it really requires expertise from people who have been dealing with them for some period of time, typically more than 10 years, and are seeing a very sufficient number of patients to maintain expertise in that particular disease.

Int: Certainly a frightening scenario for many, many people. Maureen Coleman, how do people get in touch with Carcinoid Neuroendocrine Tumour Society of Canada?

M: They can do a number of things. We have started with leaflets in hospitals, there are some doctor's offices where we have a flyer, they can also Google the name Carcinoid or Neuroendocrine and they'll find our Society. On our website there are phone numbers and email addresses, so it is harder for the patient who is not diagnosed, to find us. Once diagnosed, as long as they would be fairly knowledgeable, not even sophisticated, but just know how to use Google or a search engine and put in these words, they will find us. What would help us as well is if our medical community told their patients about us.

Int: I guess this is why we have to educate both the public at large and certainly our General Practitioners as well?

K: Certainly that appears to be a very worthwhile thing. It is a problem though that you see it occasionally; it's not going to come up to the front of their minds. And as well, for the public I think it is difficult, and I've certainly had the experience over thirty years I've been doing this, I'm sorry to say, that I see patients with much more common malignancies and I have spent a considerable amount of time explaining the ins and outs of it and what the treatment is, and I have sometimes gotten the comment saying, gee that's really

interesting, how come we didn't know about this, how come you don't broadcast this or how come you don't have it in the papers and the answer is well actually we do, but most people do not when they come to seeing this in the paper want to encounter this because it is not something that they see in a very positive light. They'd rather avoid it and you can't coerce patients or the population at large to know about this and read about it. Most of them really only find about it when they have to deal with the disease itself but otherwise there's a disincentive to actually find out about this and read about it. Only when you get it, do you really have a vested interest in finding out about it, and that's just a human problem.

M: We're finding awareness everywhere at this point. Because of NET Cancer day we're also organizing to send information to every Member of Parliament, provincial parliament in Canada about this disease.

Int: Well we want to thank both of you so much for joining us and spreading the word. Our guests have been Dr. Walter Kocha, Medical Oncologist at The London Health Sciences Centre and also Maureen Coleman, President of CNETS, that's the Carcinoid Neuroendocrine Tumour Society of Canada. Thank you both for the work you're doing. All the best to you.....