Kawasaki Disease
Protecting Tiny Hearts into the Future
An Interview with Dr Tom Johnson

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Rachael McCormack, Founder of Societi Foundation talks to adult cardioligicst, Dr Tom Johson to discuss the topic "Protecting Tiny Hearts into the Future."

Dr Tom Johnson is a top adult cardiologist, leading researcher, educator and a clinical mentor for up and coming cardiologists. He works as part of a remarkable team at Bristol Heart Institute, UH Bristol.

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Rachael: *We know from parent feedback at Societi, Tom, that families of children affected by Kawasaki Disease are really keen to know about how Kawasaki Disease might affect their child as they transition from child to adulthood. We know that long term care needs can differ depending on the severity of heart damage experienced when first ill with Kawasaki Disease. Can you talk us through what's needed and why?*

Tom: Rachael, as you acknowledge, we been running an adult clinic for Kawasaki Disease in Bristol alongside Rob Tulloh, a colleague of mine who has recently retired but with, really the expertise behind the understanding of Kawasaki Disease. That clinic came about through an acknowledgement that there is this transition from paediatric care into adulthood, which is not necessarily very well joined up. And so, often families and children -Its important to understand that there is an ongoing need for both – are lost into the adult system. That just demonstrates the fact that there is a disconnect between child healthcare and adult healthcare and there isn't, for the moment, an awareness of Kawasaki Disease into adulthood.

What's been most striking for me, running this clinic alongside Rob for the last couple of years, has been the need of both the patient and the family. Often, it's the family - the parents who have a huge number of questions and concerns, often stemming from the very first period of illness. Our clinic is evolving and the needs, I'm becoming more aware of, are fairly extensive from both the counselling around the initial treatment, or lack of treatment, or the later diagnosis, which often is something that causes a huge amount of concern quite understandably. And then our consideration is about how we are going to follow the child patient/adult into the future in terms of understanding the consequences of the initial illness.

That's evolving as we do it as well, because we're increasingly becoming aware of new needs and ongoing issues around surveillance which maybe we will touch upon later in the podcast.

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Rachael: *Thanks Tom. I know that when we've spoken before, you've mentioned that, very often, somebody could come to the adult clinic and maybe, it's a one-off visit where you're able to answer questions, reassure and provide a supportive insight into their journey of Kawasaki Disease and say, 'You know what? From this point you don't have any ongoing care needs.' And that's hugely liberating and a hugely positive outcome in terms of some of those patients. As you say, that disconnect between paediatric care and adult care leads to, in some cases, uncertainties and some anxiety. That's a hugely valuable role that the clinic also plays.*

We know that follow up care can vary a lot depending on where you are in the U.K. and too few adult cardiologists currently recognise the significance of Kawasaki Disease and the need for some patients to have lifelong care. Can you explain its importance, from your perspective?

Tom: Just touching back on the point you've made about this spectrum. That's right and, initially, it felt slightly fraudulent almost to have a clinic with well people turning up to talk through the fact that, thankfully, they haven't had persisting damage to their heart. But, on reflection, that's incredibly powerful and beneficial to both the patient and the wider family.

You're right to indicate that we have a spectrum of presentations – it may be that there was a childhood illness and there was an initial concern around cardiac involvement, but things have settled – so then the whole purpose of our meeting is to make a final reassessment of heart function, the coronary structure, take a CT scan and then simply offer reassurance. We're then just a sounding board, maybe for reexploring the initial presentation.

At the other end of the spectrum, there are patients who have persisting, significant heart involvement, particularly where we see the heart arteries having become swollen or aneurysmal as we describe it – and then there may be need for much closer surveillance.

They are the two extremes and then in between, where there has been some heart involvement, through close collaboration with leading experts, predominantly the American group in San Diego, we've generated a protocol for assessment which involves maybe a CT scan for the arteries, an MRI scan to assess both heart function and the supply that the arteries provide the heart and then that counselling element too. That may well result in annual, or two yearly, or five yearly surveillance depending on the nature of the injury that's already evident.

Rachael: *With a growing number of people affected by Kawasaki Disease in childhood, necessarily it follows, that we've got a growing cohort of those entering adulthood who have been affected by Kawasaki Disease.*

It should be on the radar of more cardiologists because that patient community is growing, unfortunately - until we can do something about the prevention of coronary artery aneurysms in particular. So, it is of significance into adulthood. Would you agree?

Tom: Yes, I would. Even in the time that we've known each other, and I've been involved with Societi, there has been an increase in awareness.

There is a danger - social media is powerful but also, social media has a risk of echo chambering, where the same people are talking to each other all the time. But certainly, within the Twitter community there are not infrequently cases of angiograms shown on our medical feeds with swollen, aneurysmal arteries and people are very quick to consider Kawasaki Disease as an underlying diagnosis.

So, on the one hand, there's an awareness and there's a willingness to consider Kawasaki Disease in what we call a differential. But we need to go one step further, which is acknowledging what that really means and how we protect for the future because there's possibly a disconnect there. OK, we see it as potentially a problem, but we don't then know what to do with it.

Within the U.K. community, I think that people know that I have an interest – there are a couple of colleagues in London that also share an interest and were involved in the writing of the extended care pathways. So, there are people to turn to, if people aren't sure but we probably just do need to broaden the educational remit here.

Rachael: *I would certainly agree, and we've taken some important steps on the journey. If we were speaking, even three years ago, we would be at the start of that route to raise awareness with more adult cardiologists and it's great to have had your support on social media and more broadly to get us into the minds of folk. I know you've spoken at a number of conferences about it and all these small steps really make a difference.*

You've mentioned coronary artery aneurysms and we know that one of the most severe impacts Kawasaki Disease can cause is giant coronary artery aneurysms that persist into adulthood and are lifelong.

I know when I first established Societi there was a prevailing view that this might be something that affected a just handful of people. We now know from our work and from your work and from research in fact, that there are many, many people who have been affected in this way. From a cardiology perspective, what are the risk factors for somebody who lives with a giant coronary artery aneurysm?

Tom: When we say giant, we are talking about a particular size – I'm not sure how many of my colleagues actually measure the size of the swellings in arteries but a threshold of 8mm is the cut-off that we use.

Where faced with that, there's quite clear guidance that we have to be aggressive in thinning the blood to prevent blood clots from forming within those swellings.

The concern there can be that the swellings alter the flow of blood, and the blood is sticky. So, if the blood stagnates or the blood pools in areas, it has a tendency to clot. The blood clots can result in a blockage in the artery which is a cause for a heart attack.

So, that's the greatest concern and a way of countering that is to thin down the blood. Historically, that's always been done with Warfarin and that's still the guidance although there are patients who use newer agents which also thin the blood. But we tend to use warfarin or a combination of warfarin and aspirin – both blood thinners.

We mentioned about this acknowledgement of the risk. The work we did in Bristol was looking at the cohorts of patients who came through our cath-lab just for normal reasons and were then found to have swellings or aneurysms. The difficulty there is, that doesn't necessarily mean Kawasaki Disease, but it could be Kawasaki Disease and so there is a whole host of patients that are potentially missed and the risk is then if you don't identify them at that time, they may come back at a later time with a clotting event – with a heart attack. That's where the education is needed, to identify and then respond to these findings with the commencement of blood thinning treatment.

Beyond that, in terms of our surveillance protocols in clinic, the reason for that is, that we know from the larger populations of Kawasaki – so San Diego, across in Southeast Asia, Japan predominantly, that there is a potential progression within the artery where narrowing can develop at both the entrance and the exit of these aneurysms or swelling areas. It can be then that the patient develops a narrowing that could then cause chest pain and could then further interrupt flow. Part of our surveillance is assessing the arteries for that type of change. That's something that the standard adult cardiologist wouldn't be so aware of – that and this risk of blood clots and the need to think about anti-coagulations. They're the sort of education areas we need to enhance and develop.

Rachael: *I would agree. I'm very alert to the fact that lifetime care is hugely important for people who have had the most serious heart damage but there is a spectrum, as you say, of patients who potentially need either follow up or that doctors need to be made aware of that prior history of Kawasaki Disease.*

When we wrote the lifetime management guidance which was published back in 2019, we included a patient specific protocol to allow documenting, by the closest clinicians to patients, of their Kawasaki Disease experience so that could be on record and referred to subsequently. Do you think that's a useful tool for clinicians?

Tom: Yes, I think its an incredibly useful tool as part of that recommendation. And now we're formulating these PSPs or person specific protocols for patients.

It's probably got as much use for the patient as it has for the caring physicians. For me, it is a unique clinic that I run. I'm not used to having teenage or young adults in clinic. It tends to be a very different demographic that I'm used to treating – people in their 50's or 60's or beyond so it's quite refreshing to have a clinic full of university graduates or students.

Often then, the PSP provides huge amounts of security. If you're moving away from home and your moving then to a new environment, a new GP or, for instance with some of the more recent patients travelling abroad for a year of study, to have a document that outlines what's gone before and the potential risks, is incredibly valuable. Within that we have our details, so that we can be contacted should there be any difficulty and acknowledging that being faced with a diagnosis you might not understand, doctors can often react poorly.

When you don't know, sometimes you just shut down communications rather than explore. It's important for there to be channels of communication open which is what the PSP offers. My details are on my patients PSP so that people can contact me should there be advice

required. It's very beneficial for both the patients and for the doctors that might end up treating them.

Part of the importance of that was an acknowledgement of the fact that these are potentially young patients who wouldn't usually be presenting with heart problems. So, in producing this letter, or protocol to a paramedic or to an emergency department if you're suffering an acute complication – so as we've mentioned the worst possible being a clot blocking one of the arteries – that you're not dismissed because you're not the 'type' to be having a heart attack, which is a potential problem.

We know that these patients can present atypically. They don't present in the usual way of a heart attack patient. One, they don't look like a heart attack patient because they are 30 or 40 years younger than they should be. They are otherwise fit and healthy and secondly, they may not have the classical symptoms of central crushing chest pains and such like. There's a risk of dispatching or overlooking because it just doesn't fit our normal diagnostic framework. That's where the PSP kicks in and clearly states 'do not overlook this patient, this patient may well be having a life threatening event, please do consider x, y and z'. So, it's there as a safety net as well as a potential instructional leaflet, if you like, for both patients and doctors who may be treating them.

Rachael: *A seemingly important tool on both sides. I can remember when we were putting the guidance together and working through the various algorithms for that atypical presentation that you describe.*

You talked about the progress that we've made in terms of raising awareness of Kawasaki Disease generally – small but positive steps with the adult cardiology community. I think there's as much to do with that recognition of the atypical presentation of a cardiac event in a young person that's had a prior history of Kawasaki Disease. A project, perhaps Tom, that we can work on in the weeks and months to come.

So, given the clear need for more cardiologists to be able to recognise the long term care needs of patients who have serious heart damage from acute Kawasaki Disease and also be alert to that atypical presentation of a major cardiac event, what guidance would you point colleagues to, if they are looking for best practice?

Tom: The document that we generated at the end of 2019/2020, available through Heart, the British Cardiac Society Journal would be the first place to look. There is some American guidance as well, but, certainly from a U.K. perspective, that would be the go-to manuscript to turn to first.

Rachael: *That is signposted on the Societi website. If you're reading our interview from a source other than the Societi website, do drop over to our website - societi.org.uk and that is prominently available and easy to access.*

Tom, thank you so much for your time today and for sharing such valued insights and your expert knowledge. I just know how well received this podcast will be with our supporters and families but also to the many, many young people and adults that are living with Kawasaki Disease today. Your support is valued very much indeed and your continued expert input into our work a charity is hugely appreciated.