Kawasaki Disease
World Heart Day 2023

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Kawasaki Disease in Adulthood - A conversation with Professor Robert Tulloh and Dr Tom Johnson

Rachael McCormack, Founder of Societi Foundation talks to adult cardiologist, Dr Tom Johnson and UK leading Kawasaki Disease expert Professor Robert Tulloh to discuss the topic "Kawasaki Disease in Adulthood."

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 clinical team in Bristol and is a member of the Societi Scientic Advisory Board.

Professor Robert Tulloh is a paediatric cardiologist and world recognised specialist in Kawasaki Disease. Prof Tulloh is also a Societi Foundation Trustee, and has helped guide the work of our charity since it's inception in 2015.

Rachael: *Hello and welcome to our World Heart Day podcast series focused on the impact of Kawasaki Disease into adulthood, and its relevance to adult cardiologists. I'm Rachael McCormack, Founder of Societi, the UK Foundation for Kawasaki Disease.*

I'm delighted to be joined by Dr. Tom Johnson, Associate Professor of Cardiology in Bristol. Welcome, Tom to our conversation today.

Tom: Thanks, Rachael. It's lovely to be with you.

Rachael: *Tom, you've been a member of the Societi Foundation Scientific Advisory Board for a number of years now. You've been an important expert contributor for us. And as an adult cardiologist, really key to this advisory group. You have a special interest in Kawasaki Disease and you've been an active researcher in this area as well. And of course, a prominent ambassador for the need for more adult cardiologist to get familiar with the serious long term heart damage that Kawasaki Disease can cause. Tom, you also lead a long term follow up clinic for adults by referral for those affected by Kawasaki Disease in the UK. Can you tell us a little bit about that?*

Tom: Sure. So, the interest with Kawasaki Disease really stemmed from collaboration with my colleague in Bristol from the paediatric side, Professor Rob Tulloh who has a national and international reputation in terms of the management of Kawasaki Disease, and I very much piggybacked on to that where interventional cardiology, which is my sub-specialty interest within cardiology, has an interesting role, particularly for patients who have had Kawasaki Disease with coronary involvement.

So I've come at this from a slightly different angle. Rob's now retired and so I'm managing these patients on my own, where actually the decision making is quite challenging, and hopefully today we can discuss some of those challenges. I think both in the transition of care from paediatrics where a diagnosis of Kawasaki Disease has been made previously, and how those patients transition into adult services and ongoing follow up.

Actually the area, that for me, I think is even more challenging are the adult patients who present without a history of Kawasaki Disease, but with clear evidence of coronary involvement that has led to a presentation, for instance of heart attack, and where we, as the adult cardiologists really need to have a better focus on detecting those abnormalities and reacting to them. So I think there are two different issues that are actually come about through this very useful collaboration and partnership that Rob and I've built up in Bristol.

Rachael: *Thank you, Tom. I'm keen to get into that conversation. But before we do, I'm delighted that Professor Tulloh has been able to join us as well today. Rob, you're a founding member of Societi, Chair of our Board of Trustees, and a leading voice as Tom has mentioned in Kawasaki Disease for many decades. And perhaps seeing more children with Kawasaki Disease over your career before retirement than any other clinician in the UK. You're also a prolific researcher in the field. So thank you very much for joining us.*

Rob: Thank you, Rachael. It's good to be joining you and Tom today in this important conversation, and I look forward to helping to spread the message about the importance of Kawasaki Disease.

Rachael: *Thanks again for joining us, Rob. So turning to our conversation topic for today, Kawasaki Disease into adulthood. Tom, can you talk us through some of the issues that present?*

Tom: So I think as an adult cardiologist, I'm not necessarily best placed for describing the consequences of Kawasaki Disease in infancy. But in terms of this transition of care

Victoria Court, Holme Lane, Winthorpe, Newark, Notts. NG24 2NU
email: info@societi.co.uk web: societi.org.uk

– certainly our lifetime guideline hangs upon the presence or absence of coronary involvement. And clearly that is dependent – for adult cardiologists who may not have an understanding of this – that is dependent upon the early diagnosis at the point of presentation as infant or child and then the delivery of treatment being immunoglobulin therapy.

Now we know that of those children presenting with Kawasaki Disease with early treatment, 20/30% may have persisting coronary involvement. The difficulty – and what Societi has done such a good job in terms of raising awareness – is about trying to overcome this gap that we currently have of failure of diagnosis and then not getting treatment where then the risk of coronary involvement is incrementally higher. So, for us in the adult service, our ongoing care is very much driven by the presence or absence of coronary involvement. If the coronaries are no longer involved and actually it may be a single assessment and discharge from our service. But where coronaries are involved from ectasia, through moderate aneurysm to giant aneurysm, then actually there is a requirement for ongoing assessment on an annual basis with MRI, cardiac CT. And I have to admit, some of that is of academic interest and we're learning I think, in terms of how we develop these pathways. And it offers a challenge but also quite a lot of interest, I think, for us as an adult cardiology community.

Rachael: *So you've mentioned where we've got children who've had a known prior history of Kawasaki Disease. And in this podcast series, we hear from Kris who's in his 30s. He was affected by Kawasaki Disease at the age of 10. Diagnosis was delayed and that's often the story with older children. He went on to have some fairly significant heart damage. So for a patient joining you in clinic, what are the considerations, Tom for you as a cardiologist.*

Tom: So I have to say that the patients presenting to clinic with a prior history of Kawasaki Disease offer different challenges to those where the diagnosis hadn't previously been considered. And that often is due to the interesting scenario for me as an adult cardiologist, where I'm often faced with both my patient having transitioned from paediatric care into adulthood alongside their parent, or parents. And, there are then a couple of issues that we have to address. One is the ongoing management of the coronary abnormalities, and the other then is unpicking some of the obvious distress and anxiety that has associated with the treatment through childhood. That's often parents actually grappling with a feeling that they may have not reacted soon enough and often then anger also to the medical teams for having not responded or delivered care in a timely fashion. So the fascinating thing about developing this clinic alongside Rob was actually learning new skills and communication – with not just your patient, but associated caregivers or family members. And so it does make for a very different type of clinic experience to my normal general cardiology service.

Rachael: *Thank you Tom. To Rob, if I can just bring you in. Tom's touched on the transition from paediatrics services into adult care and the importance of the handover as well as active role for the adult cardiologist, perhaps alongside paediatrics. What do you think "good" looks like in terms of that transition?*

Rob: I think that's a really interesting point. The children who have had Kawasaki Disease and coronary artery involvement have usually been very invested along with their families in terms of medical care. They are very clued up as to what's going on. The paediatrics and the paediatric cardiology clinics are well familiar with Kawasaki Diseases now. Previously, it used to be thought to be a very rare disease, but people are realising now how much more common it is, than has been previously thought. Therefore, good information, good communication and good engagement is really important, especially as the child turns into an adolescent. We know that adolescents are not very good about engaging with healthcare services, about taking their medication. But somehow we need to facilitate the transition into adult care. And this is where it's been so valuable working with an expert such as Tom, who really understands the coronary arteries probably far better than I do, even though I understand Kawasaki Disease. And so I've had a great learning experience as well. And this has allowed the child to become an adult, to take responsibility and engagement for their own disease for the first time. And to gradually to have it handed over from the paediatricians pick a paediatric cardiologist, and from the parents, to the child themselves and to adult cardiologists. So that when there are problems in the future, they are much more engaged and they don't dismiss it. And this helps with the understanding and the communication for the future.

Rachael: *Thanks, Rob. That's a hugely helpful insight and you've mentioned increasing incidence, so these hand overs are going to need to take place more and more frequently. And we've heard from Tom, just a few moments ago about*

the challenges of that process of transition for a child who's had a prior diagnosis of Kawasaki Disease. But, what about in the situation where there is a presentation of a patient and you perhaps have no prior knowledge of Kawasaki Disease?

Tom: Yeah, so I think we have a duty of care as an interventional cardiologist undertaking coronary assessments - so an invasive coronary angiogram and both reporting and reacting to abnormalities in the coronary vasculature. So actually, with Rob, we undertook an analysis in Bristol of our patients under the age of 50, presenting through the cath lab and interrogated every angiogram, and found actually a fairly significant incidence of ectasia, and aneurysm - and more concerningly, that that was seen much more commonly in the under 50 year old population presenting with acute coronary syndrome presenting with either ST elevation or non ST elevation infarct. So the message there being that coronary involvement from Kawasaki Disease, places you at a much higher risk of a thrombotic event of obstructing an artery. And obviously, we react in that sense to treat what's in front of us. But actually there is an opportunity to reduce risk, potentially to acknowledge these changes before a patient comes to harm and to then react by anticoagulation. Or at the time of reacting to a heart attack, then ensuring that ongoing risk is addressed by formal anticoagulation. The guidelines would stress Warfarin in combination with aspirin. And so really, it's ensuring that point of potential diagnosis is not missed.

Rachael: *And that's actually a point that a number of the patients who we've interviewed as part of our podcast series have, really emphasized. I think, being a young adult with a very complicated heart history has presented challenges in some areas for them in just engaging with services and clinicians in turn engaging with them. And so that's a really important point on to underline. That just about brings our world heart a podcast on Kawasaki Disease into adulthood to a close, but I will pass back to Rob and then to you, Tom - just for any closing thoughts that you might have. Firstly, Rob.*

Rob: Yes, I think it's an interesting discussion. And I think that paediatricians certainly need to be aware that children who are older than 2/3/4/5 years old, can get Kawasaki Disease and in fact, we found that about a quarter of our patients were outside that age group. So think of a child who maybe 10 or 12 could be presenting with Kawasaki Disease, if they have the classic symptoms. And these children often present later. They are diagnosed later and treated later. And they may run the risk of having worse complications as a result. It's important also, to remember that it's not just a self-limiting disease that goes away and causes no long term problems. We know and we see many children who become adults who do have ongoing cardiac complications, and need proper transition and proper long term management and into the adult clinics.

Rachael: *Thanks Rob, it's really helpful to have your input and your insights today. And, over to you Tom, any closing thoughts from you?*

Tom: Well, I think as I've probably already stressed the importance of us, recognising coronary abnormalities, in the absence of a clear cut history of Kawasaki Disease, is for me, I think where we're likely to have the greatest impact initially. Whilst the awareness of Kawasaki Disease increases, and there is undoubtedly going to be a demand in terms of this transition from paediatric to adult care. But at the same time, although I've stressed the importance of acknowledging aneurismal or ectatic disease and the need to anti-coagulate, I think then there's this need to understand that there is then a lifetime management.

We haven't talked about person specific protocols. We haven't talked about the ongoing management in terms of annual surveillance. There's a lot more to consider in terms of the ongoing management of these patients. You call me an expert, I don't feel like an expert. I feel like I'm an interested physician, who is just trying to grapple with a long term pathway of care. And so I think the more that we acknowledge that we have more to learn, the more exciting it is, as a clinician, that actually probably the more impact, we're going to have long term for a group of patients that are really a joy to engage with.

Rachael: *Thank you, Tom. You've given me a brilliant opportunity there to invite you both back to do a subsequent podcast. And we'll look at some of those other issues that we haven't been able to explore today. But thank you again.*

Thank you for listening to our World Heart Day podcast series. You'll find a whole range of podcasts linked to this episode here on SoundCloud. I hope you enjoy listening to them. And my sincere thanks again to our fantastic speakers today, Dr. Tom Johnson and Professor Robert Tulloh - Thank you gentlemen, very much indeed.

Victoria Court, Holme Lane, Winthorpe, Newark, Notts. NG24 2NU
email: info@societi.co.uk web: societi.org.uk