

#### Rachael McCormack, Founder of Societi Foundation talks to Dr Damian Roland to discuss the topic "What everyone needs to know about Kawasaki Disease."



Dr Damian Roland is a Fellow of the Royal College of Paediatrics and Child Health, an active researcher in children's medicine, a clinical educator, a leadership mentor and is hugely passionate about child health and sharing knowledge.

**Rachael:** As an emergency medicine consultant working in a busy paediatric emergency department, what are you on the lookout for in children presenting at A&E with suspected Kawasaki Disease? What is it that you notice first? What is it that makes you think 'this feels like Kawasaki Disease'?

**Damian:** There is one thing that really stands out that does differentiate Kawasaki Disease from virtually all other illnesses that children can get, and it is the degree of irritability that they have. Irritability is a funny word but what I am meaning here is, when children have a bit of a fever, we can describe them as poorly, maybe a bit miserable - a bit sorry for themselves. But when I use the term irritability, it is almost an anger inside that child. They can never be still. One of the interesting characteristics, and this doesn't apply to all children, but a lot - is most children, when they have a bit of a cough and a cold, Mum and Dad can put their arms around them, and they can be comforted by them. Children that are irritable with Kawasaki Disease sometimes don't even like that. Even a hug is painful to them and they will sometimes sit with a funny posture because their joints are hurting. That does stand out differently from other children with coughs and colds. I mention the parents experience too; you are often looking for the subtle cues that parents give you that this is not a typical illness. Parents know their children very well, they're likely to have experienced lots of coughs and colds in the past but there is something different about this illness. They can't console their child and there is something much worse about them in the way they are feeling than there is in other diseases.

**Rachael:** As a parent of a child who has had Kawasaki Disease, I recognise that, and I remember really vividly holding my daughter in the middle of the night and her just wriggling and squealing and just not being able to be consoled. So, as you say not a traditional, clinical sign or symptom of an illness, but that irritability does bring something for doctors to be alert to and parents to be aware of. So, beyond that non-specific irritability what else are you looking for in a child with Kawasaki Disease?

**Damian:** If we are talking traditionally (textbook) there are some criteria that we use to diagnose Kawasaki Disease with. One of those is, it is not just a couple of days of fever, it should be 5 days of fever and there are a couple of specific features these children have. And one of the challenges of Kawasaki Disease is these features aren't specific just to Kawasaki Disease, there are other illnesses that present with this. It is just the constellation of these symptoms coming together.

So, what we are looking at is; 5 days of fever plus bilateral conjunctivitis and I've emphasised the bilateral. It is not just one bunged up eye. It is both eyes that are red and streaming. You also have, the medical term being, cervical lymphnodes (basically swollen glands), cracked lips or strawberrytongue and possible peeling of the hands and feet so you can see little bits of the skin coming off.

One of the challenges with that particular sign is that it is quite late in the disease process, and you really do need to be moving forward in the diagnosis if that feature is present. The last thing which is very non-specific is polymorphous exanthem. Polymorphous means everywhere and exanthem means rash. So in amongst all the children I see in my emergency department who simply have a fever or a cold, what I am looking for is the child with the prolonged fever and has at least 4 out of the 5 features that I just described. Now one of the challenges is as research of Kawasaki Disease has progressed is that we know that sometimes we don't get to the 5 days of fever, but you clearly, obviously have those symptoms. And sometimes you may not have all of the features mentioned but your suspicion is so high that you are sure it is Kawasaki Disease and you will jump in anyway.



**Rachael:** *And in fact, relatively recent NICE guidance has encouraged doctors to think about Kawasaki Disease with persistent fever and there may be no other features of very few of those features and I think that responds to research that shows quite clearly that it is often the youngest babies and children with the fewest features that have the poorest outcome. So, there is a certain conundrum. Kawasaki Disease is nothing if not enigmatic, but it certainly presents a challenge to Doctors.*

*We often hear from families who have waited a long time before getting a correct diagnosis. Perhaps that is a result of that varied clinical presentation, which can lead to delayed treatment of Kawasaki Disease and consequences for our children. As a consultant in a busy paediatric emergency department, you see a lot of children with so many different conditions. So, why in your view is Kawasaki Disease so hard to diagnose?*

**Damian:** *I think there's two reasons. The amount of children presenting to urgent and emergency care is quite staggering. For example, in my department yesterday we saw over 200 children and 40% or more of those children will have had a fever. A percentage of those children will have appeared quite irritable, some of them will have had rashes, lots of them will have had raised lymph nodes – but none of them had Kawasaki Disease. So, there is a problem of scale here and it isn't easy to pick out because there's such a melee of children to deal with.*

*On top of that problem, you need to be thinking about something to be able to rule it out. I think this is one of the great things you've done with the charity – if you're not aware of the problem, why would you ever think about it? Everyone knows about the common cold, everyone knows about pneumonia, even lay people know about urinary tract infections, so you think of those things early on in your diagnostic profiling. But even doctors, especially those who are new to paediatrics, or haven't worked in child health for a long time, may never have even heard of Kawasaki Disease. Or they heard of it at medical school and then it's just simply slipped away from them. So, it's a double headed challenge – making sure that people are aware of it to think about it but also it is really difficult in the landscape of the current emergency and urgent care system.*



**Rachael:** *We know as a charity raising awareness of Kawasaki Disease, that awareness point is absolutely crucial. It's difficult to think of something if you're not familiar with it. And perhaps worse, as another side to it, if you were made aware of it through information that was available maybe 10 years ago, maybe current knowledge and current incidence isn't at the uppermost of your thinking, but we know Kawasaki Disease well as a serious illness. As a leading expert in children's emergency medicine, you see many serious conditions every day which need rapid treatment to make sure children have the best possible outcome. From your perspective, why is Kawasaki Disease a medical emergency?*

**Damian:** *It's a really good question because the concept of what is a medical emergency, I suspect, is very different whether you're a healthcare professional or a parent. One of the challenges of medicine is making sure that we're looking at the same outcomes. For an emergency department doctor or a paediatrician, you're looking at the unwell child in front of you and trying to treat that unwell child. But the consequences of Kawasaki Disease, especially for that group of children who end up with heart problems as a result, it is lifelong. I think that's one of the criticalities of Kawasaki Disease, is that once we have recognised and we think this is what the disease is, the treatment is really important because we can reduce that lifelong morbidity if we act soon enough. That's one of the really important points about this particular disease, is its longer term impact, especially if missed.*

**Rachael:** *And there about a quarter of all children affected by Kawasaki Disease who will have some heart involvement and about one fifth affected will go on to have lifelong heart damage, so that point is really well made in terms of needing a call to arms and urgent action if Kawasaki Disease is suspected.*

*Damian, thank you for your time today to share your valued insights and expert knowledge and understanding of Kawasaki Disease. In our work to get this sinister disease known, your support is very much valued and your continued expert input into our work is hugely appreciated. Thank you.*

