

Rachael McCormack, Founder of Societi Foundation talks to Professor Robert Tulloh about Kawasaki Disease to present some important facts and help to dispel the myths.



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 its inception in 2015.



Rachael: *Rob – As a prolific researcher in Kawasaki Disease over time, can you tell us a bit about your work over the last 2 or 3 decades on Kawasaki Disease — what do we know now that we didn't know in the 80's – and what is still to be learned about Kawasaki Disease?*

Rob: Thank you Rachael. This is indeed an exciting time to be involved in Kawasaki Disease, since we are making great progress and helping to treat it better.

In the 1980's it was indeed a rare disease, but it was known to have an important list of symptoms. These are a fever which lasts for 5 days or more. Also – other symptoms of soreness which as red eyes (which are not sticky), red mouth and cracked lips, red or swollen fingers, a rash over the chest and large swollen glands in the neck. Studies in America and Japan led to the trials showing that Immunoglobulin (or Antibodies) given by drip and early in the acute disease, made the heart damage less bad. Since then, high dose aspirin has been added to the acute treatment to damp down the soreness and low dose aspirin is carried on until doctors know that the heart has not been affected.

Over the last 30 years, many other facts have come to light, whereas in some areas we are still unsure. We have learnt that there is an active immune response. It is not that people have weak immune systems but that they respond badly to an infection or toxin. Therefore, all the initial treatment is about damping down this immune response, and there are other second line agents that we can now use – such as infliximab, anakinra and tocilizumab. We also think that steroids should be good treatment – but studies have not been clear on this point. So, we are doing a large trial across the whole of Europe, to see if we can prove whether they work.

We have looked at cases that we are seeing and it is clear that the case numbers are rising fast across the world. It is roughly 10 times more common than it was 30 years ago. It is the most common condition in the UK to affect children's hearts after they are born – now more common than scarlet fever, and even more common than some types of meningitis. Meningitis is well known, but many people have not heard about Kawasaki Disease and some doctors don't seem to be able to diagnose it.

We have done a large research study in the UK and Ireland of every case, and this has shown how bad we are at picking up and treating this disease. The earlier it is treated, the less the damage to children's hearts. A quarter of all children will have some heart involvement from Kawasaki Disease, which is why I see so many cases as a heart doctor. We now know that even minor changes in the heart can lead to lifelong consequences. We have had agreement from GPs, A+E doctors and children's doctors as to how this disease should be treated.



Rachael: *Rob, given the work that has gone on in recent years, can you tell us why are there so many myths and so much misinformation out there, on Kawasaki Disease?*

Rob: There are two main problems with doctors treating Kawasaki Disease early. The first problem is that the first symptoms of a fever and rash can occur with many childhood infections. We certainly would not want people to think that every childhood fever is Kawasaki Disease, this would cause panic and not be helpful. However, a child who has a cold will develop a runny nose or a cough within a couple of days of having a fever. These are not symptoms of Kawasaki Disease. Or they may have diarrhoea – which is not a major symptom of Kawasaki Disease. However, if 4 days have gone by, with a fever, rash, and red eyes, sore mouth, irritability and no other symptoms, and especially in a young baby, then the child should be suspected of having Kawasaki Disease and a children's doctor or senior A+E doctor should see them. It is important to remember that the fewer the number of symptoms, then the worse the disease and heart damage.

The other problem leading to myths about Kawasaki Disease is with doctors and nurses who are out of date. They still think it is a rare condition. This is simply not true. There are over 1000 cases of Kawasaki Disease every year in the UK. This is not a rare condition and every year I see children in

whom the diagnosis has been missed – or treated late, for which there may be a bad outcome. Early treatment at 5 days – will really help to avoid long term damage to the heart.



Rachael: *Families understandably want to know what is the cause of Kawasaki Disease. What can you tell us about the possible cause and is there any recent research that might help families understand more about this disease?*

Rob: Strangely, we do not yet know the cause of Kawasaki Disease. Doctor Jane Burns in America has shown that some people may have the genes which make Kawasaki Disease worse. She has also shown that there is something in the air that makes outbreaks of Kawasaki Disease more likely. However, no research has yet given us the cause, so that there is still no proper test that a doctor can apply in order to diagnose Kawasaki Disease. It will be found, but not yet.



Rachael; *What are the most common myths you hear about Kawasaki Disease? And can you dispel them for us?*

Rob: The common myths are that

1. Kawasaki Disease is rare. It is not. Every hospital sees a case every week or month, but often doesn't recognise it.
2. Because it is thought to be rare, It must be something else, such as slapped cheek (which is due to a well-known virus) or rheumatic fever (which is now rare in the UK). Often doctors and nurses forget about the more common Kawasaki Disease.
3. Another myth is that there is no treatment, or that the treatment can be given at any time. This is simply not true. We have good treatment and the earlier it is given, the less damage there will be to little children's hearts.
4. People believe that Kawasaki Disease does not matter and just burns itself out. This is also not true. Luckily, for some children, there is no damage and no long-lasting effect. However, for others, there is severe damage to the heart which can be life threatening. You cannot tell at the start of the illness, which this will be – so it must be treated early before it is too late.

5. Another myth is that Kawasaki Disease is only seen in children between 1 and 5 years old. This is definitely not true. The worst cases are often seen in the youngest babies at around 2 or 3 months old. But Kawasaki Disease can also be seen in children older than 5 years, or teenagers or even in adults.

5. Some people believe that a high fever in a young baby is due to a cold. This is very unlikely. In fact, the guidelines say that a high and fever over several days in a baby with no other symptoms is most likely to be due to Kawasaki Disease.

6. The last myth might be that the more symptoms of Kawasaki Disease that a child has, then the more likely he has Kawasaki Disease or heart disease. As we said above, this is not true. A baby with just a fever and rash is the most likely to have the worst heart damage and needs to be treated early.

There are many other myths and I am more than happy to discuss these with doctors around the country who want to know more about Kawasaki Disease.



Rachael: *What are the important facts about Kawasaki Disease all doctors/ parents/carers should know?*

Rob: It is important that doctors/ parents or carers remember that most fevers are not due to Kawasaki Disease. We would not want to cause unnecessary panic. However, a fever should not last for 5 days without an obvious explanation, such as a cold, or ear infection or something with a clear diagnosis. If there is no obvious cause, then Kawasaki Disease must be considered. It is a treatable condition and the earlier it is treated, then the less the risk of heart damage for the long term.



Rachael: *We are incredibly grateful to you for taking the time to talk to us today, for your continued support and determination to help get Kawasaki Disease known. If you've enjoyed listening to this Societi podcast, check out the other three podcasts in our World Heart Day series and help us get Kawasaki Disease known.*