WINTER 2023-2024

KAWASAKI DISEASE RESEARCH CENTER

Rady Children's Hospital and UC San Diego

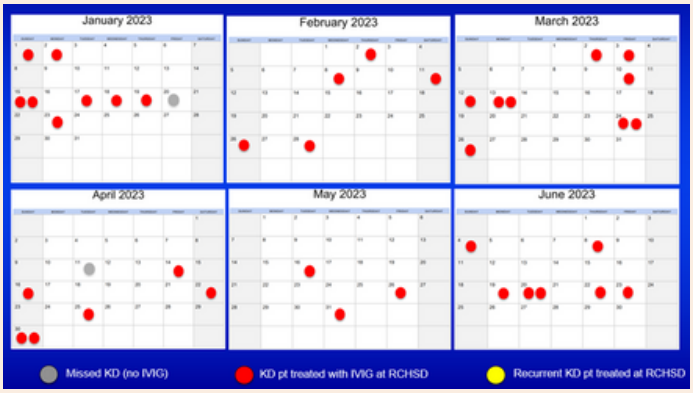
WHAT'S INSIDE THIS ISSUE:

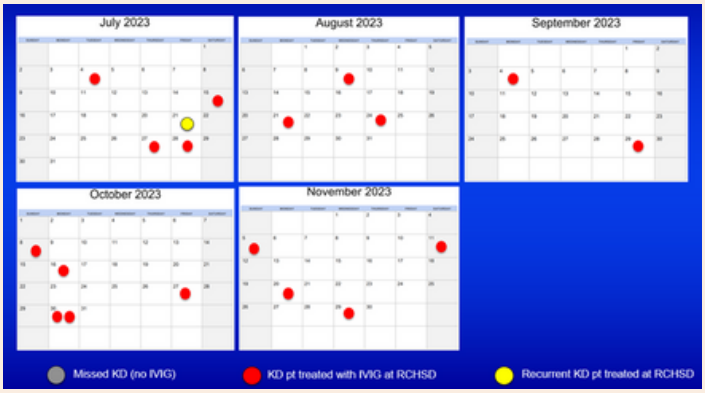
Dear KD Families & Friends,

First and foremost, the team at the Kawasaki Disease Research Center hopes that this newsletter reaches you and your loved ones in good health and that you will enjoy some cozy together time over the holidays. Our research continues to reveal new insights into Kawasaki Disease and we want to share some of the excitement with you!

We invite you to join us in Montreal, Canada on August 26-29, 2024, for the 14th International Kawasaki Disease Symposium for a chance to hear the latest about KD research and to meet with the world's KD experts. Dr. Adriana Tremoulet, Associate Director of our UCSD/RCHSD KD Research Center, is the co-president of this meeting. (Refer to page 5 for more info on how to join us.)

KD Cases in 2023: Each red dot is the onset date of a new KD case





SUBGROUPS OF KD PATIENTS

Dr. Hao Wang published his analysis of over 1,000 KD patients from RCHAS using a machine learning approach that identified subgroups of KD patients. Within each subgroup, patients shared clinical and laboratory characteristic and had similar disease outcomes.

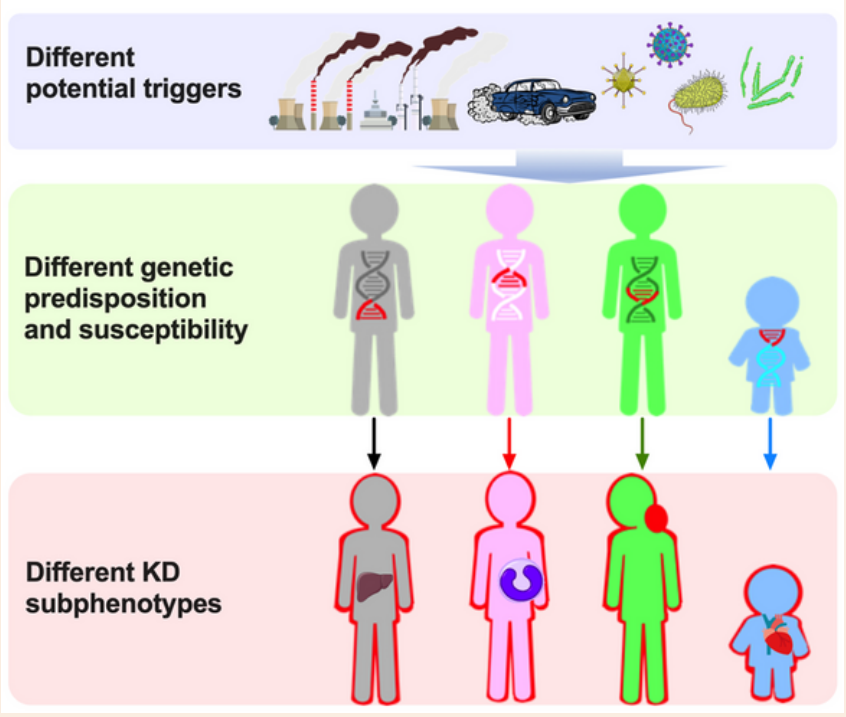
This novel analysis suggests that there may be different triggers for KD that affect children with slightly different genetic susceptibility patterns. Resistance to IVIG infusion and coronary artery aneurysms (CAA) were distributed differently across the 4 groups.

Lancet Child Adolescent Health **2023 Oct;7(10):697-707. PMID:37598693**

Subgroups of children with Kawasaki disease: a data-driven cluster analysis

Hao Wang, Chisato Shimizu, Emelia Bainto, Shea Hamilton, Heather R Jackson, Diego Estrada-Rivadeneyra, Myrsini Kaforou, Michael Levin, Joan M Pancheri, Kirsten B Dummer, Adriana H Tremoulet, Jane C Burns

Click here to read the article



Proposed Kawasaki Disease Model: Interactions between different environmental triggers and genetic susceptibility lead to activation of a common disease pathway that varies among patients and results in discrete clinical subgroups.

Direct oral anticoagulants (DOAC)

Dr. Kirsten Dummer summarized our experience with the new direct oral anticoagulants (DOACs) for KD patients with giant aneurysms who need systemic anticoagulation (blood thinners). The experience was favorable among the 24 children and adults treated with DOACs.

We have now switched almost all of our giant aneurysm patients over to apixaban so that they now take a pill by mouth twice a day instead of a shot. We will continue to collect data about outcomes in this patient population, but all the experience to date has been favorable.

***JAMA Network Open*. 2023;6(11):e2343801.**

Research Letter | Pediatrics

DOACs in Patients With Giant Coronary Artery Aneurysms After Kawasaki Disease

Kirsten B. Dummer, MD; Koichi Miyata, MD; Chisato Shimizu, MD; Adriana H. Tremoulet, MD, MAS; Jill Gleason, RN; John B. Gordon, MD; Jane C. Burns, MD

Click here to read the article

Infliximab comparison study

Dr. Koichi Miyata analyzed data from our KD patients treated with different regimens over the last 2 decades and learned that early treatment with the monoclonal antibody Infliximab resulted in a greater likelihood of aneurysm regression. This is important information that is guiding our treatment of patients with dilation or aneurysms seen on their first echocardiogram.

***Miyata K, et al. Arch Dis Child* 2023;0:1–6.**

Infliximab for intensification of primary therapy for patients with Kawasaki disease and coronary artery aneurysms at diagnosis

Koichi Miyata ,[1] Emelia V Bainto,[1] Xiaoying Sun,[2] Sonia Jain,[2] Kirsten B Dummer,[1,3] Jane C Burns,[1,3] Adriana H Tremoulet[1,3]

Click here to read the article

15TH ANNUAL KD PARENT SYMPOSIUM

WEBINAR EVENT

KAWASAKI DISEASE RESEARCH CENTER
RADY CHILDREN'S HOSPITAL/UCSD

This year the symposium was held as a remote event in November 2023 with 135 participants from 20 different countries joining through live-streaming. We were delighted to have Lucia Acosta, one of our KD parents as our Spanish translator.

The recording of the live-stream webinar has had almost 325 views since the symposium. A huge thank you to the KD Foundation in Boston for facilitating the live-streaming and supporting the event!

To access the webinar recording use the link below:

15th Annual KD Parent Symposium Link

**You may find the three flyers mentioned during Dr. Burns' presentation (late features of KD, Z-scores, KD exit form) on page 8-10.*

JOIN US AT THE 14TH INTERNATIONAL KD SYMPOSIUM & PARENT MEETING

Montreal, Canada; August 26-29, 2024

Join KD families from around the globe and listen to the latest research about KD. There will be special sessions for KD families to meet with KD global experts. **REGISTER TODAY** so you don't miss the early bird registration deadline on March 31, 2024.

Two ways to register:

1. Click **here** to register

OR

2. Scan the QR code

For additional information about IKDS go to **ikds.org** and subscribe to the IKDS newsletter.

We look forward to seeing you in Montreal in August 2024!

JOIN TODAY!

ADULT KAWASAKI DISEASE STUDY

LONGITUDINAL STUDY ON LONG-TERM EFFECTS OF KD

WILL MY CHILDREN HAVE KD IF I HAD IT?

AM I AT HIGHER RISK FOR CARDIOVASCULAR DISEASE?

WHAT ARE THE LONG-TERM EFFECTS OF KAWASAKI DISEASE?

At the KDRC, thanks to funding and support from the Gordon and Marilyn Macklin foundation, our team of researchers continues to work toward a greater understanding about the short- and long-term health outcomes of Kawasaki Disease patients through the AKD study.

If you or a loved one had KD in childhood and would like to help our team gain more insight into the long-term effects of KD in adults, we invite **YOU** to participate in the study!

To learn more about the study and how to participate, email us at **adultkd@health.ucsd.edu**. We hope you join the study today and become part of something bigger!

Visit the AKD Website

Follow us on Instagram

We are so grateful to the wonderful families whose donations have made all the research progress possible. Our challenge going forward is to continue to support our important research. Our team at the KDRC relies entirely on grant support and donations to understand the disease and improve outcomes for KD patients here in San Diego and worldwide.

We hope you will consider a gift to support KD research this holiday season.

1. Click here to donate.
2. Select “Other” in the Gift Designation drop down box.
3. Type “KD research” in the text box labeled “Designation of your choice”.

Thank You

We hope you have found this newsletter informative. If there are topics you would like to see covered or questions that you have, please contact us at kdresearch@ucsd.edu and we will include answers to your questions in our next newsletter.

Wishing you quiet moments of reflection and joyous time with your family over the holiday season.

With warmest regards,
The KDRC Team

KAWASAKI DISEASE RESEARCH CENTER
RADY CHILDREN'S HOSPITAL/UCSD

Late features of Kawasaki Disease

Peeling of fingers or toes

(Periungual Desquamation)

Keep an eye out for peeling on your child's fingertips and toes! Patients with Kawasaki disease may develop this type of thick peeling around the fingernails or toenails, usually two weeks after the start of their illness.

Nail indentations

(Beau's Lines)

Search for horizontal nail grooves appearing six weeks after the onset of Kawasaki disease.

If your child develops **peeling** under the nailbed or **grooves across the nail** following an illness that involved **fever, rash,** and **red eyes**, please contact your doctor and ask about Kawasaki disease. If there are concerns for missed Kawasaki disease, your doctor can contact the hospital page operator at 858-576-1700, ext. 0 between 9am and 5pm and ask to speak to the Kawasaki disease doctor on call.

To learn more about Kawasaki disease, watch **"Kawasaki disease: A Parent Guide"** on YouTube and visit the KD Research Center website at **https://medschool.ucsd.edu/som/pediatrics/research/centers/kawasaki-disease**

I was diagnosed with Kawasaki Disease when I was __ years old.

What is Kawasaki Disease?

Kawasaki Disease (KD) is an inflammatory disorder that can affect the heart and the coronary arteries. Some signs of KD are:

- Fever

Bloodshot eyes

Rash

Red lips and tongue

Swollen lymph nodes in the neck

Swollen hands and feet

When left untreated, up to 25% of patients can develop swelling of the small arteries in the heart named coronary arteries. If the swelling is significant (aneurysm), there is a risk of a blood clot and heart attack.

Echocardiogram results

Z-scores of the coronary arteries indicate the size of your coronary artery compared to normal values based on body size. For those who have a persistently abnormal Z-score, it is important to follow up with an adult cardiologist after your last visit in KD clinic at Rady Children's Hospital.

Z-Score	Evaluation
< 2	Normal
2.0 - 2.4	Dilated
2.5 – 4.9	Small aneurysm
5.0 – 9.9	Large Aneurysm
> 10	Giant Aneurysm

My Z–scores:

☐ My Z-scores were always in the normal range
☐ My Z-scores were dilated but resolved.
Max : _____
☐ My Z-scores are persistently abnormal.
Max : _____ Current : _____

Other changes on the echo

☐ Patent Foramen Ovale (PFO) is a natural hole between the top chambers of the heart. About 20% of people still have a PFO as an adult. If you have a PFO, you should avoid deep scuba diving and discuss with your PCP.

Things I should know

Things to consider for heart healthy lifestyle

Good for Heart

- Exercise
- Maintain ideal weight for height :

Current percentile for height : _____
weight : _____

- Maintain normal blood pressure :

My blood pressure today : _____

- Maintain a heart healthy diet
- Maintain normal blood values for lipids

My recent total cholesterol, HDL, and LDL value :
Date : _____
Cholesterol : _____ (Heart Healthy : < 200)
HDL :_____ (Heart healthy : > 60)
LDL : _____ (Heart healthy : <100)

Avoidance of the following are very important to keep your heart healthy:

- Tobacco/Cigarettes
- Amphetamines
- Cocaine

Medications for my heart recommended by my Kawasaki Doctor:

☐ None
☐ Aspirin
☐ Plavix
☐ Statin _____
☐ Direct oral anticoagulant _____

Why did I have Kawasaki Disease?

You had Kawasaki Disease because:

1. You have a pattern of genes which made you susceptible to KD and
2. You breathed in something from the air that triggered KD (the exact agent is not yet known)

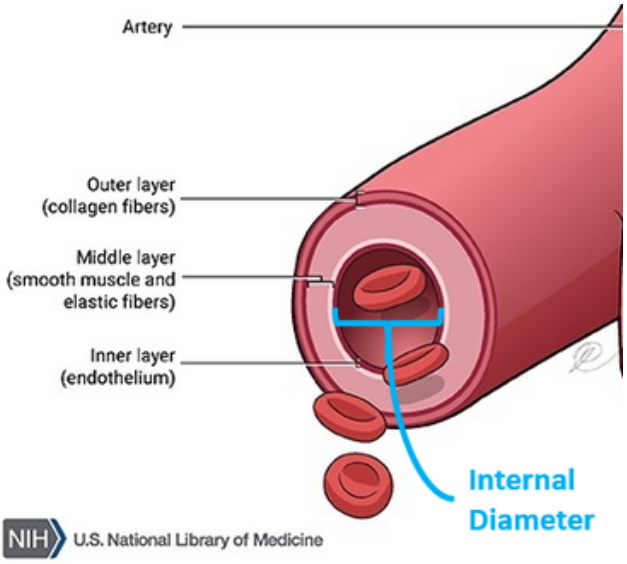
Because you carry the genetic pattern for KD, if you have children in the future, they may inherit this pattern and be at slightly greater risk of developing KD.

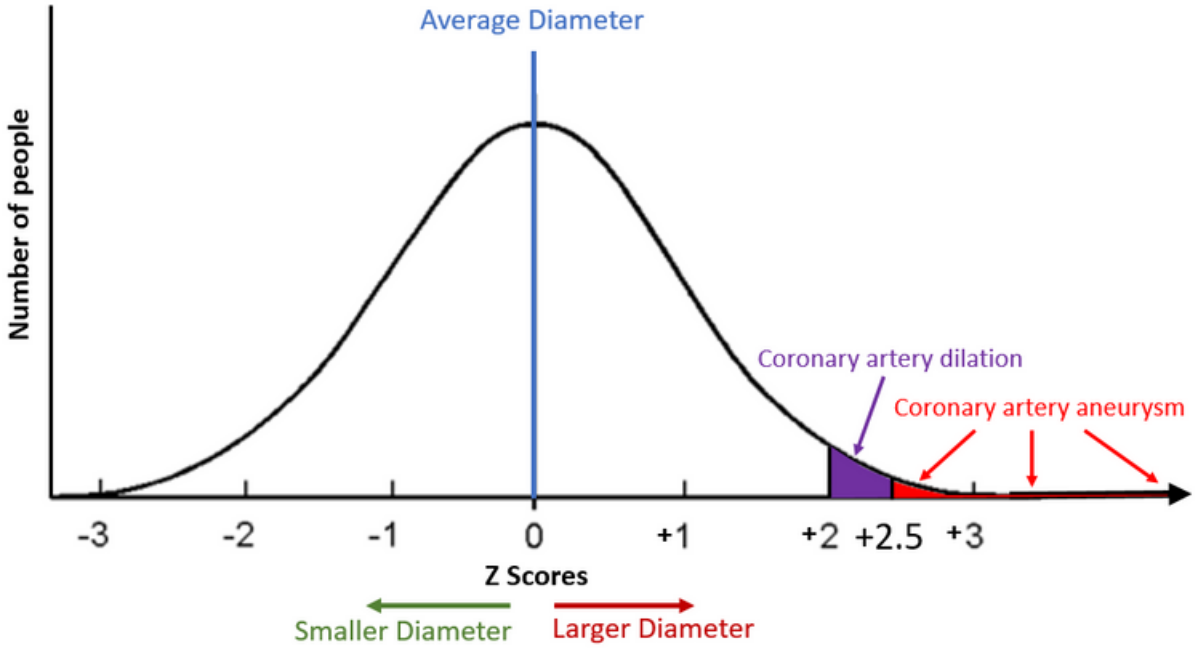
Kawasaki Disease: Z Scores

WHAT IS A Z SCORE?

A Z score normalized for body surface area represents how much larger (or smaller) a *measured* coronary artery internal diameter is compared to the average coronary artery diameter for a child of the same size (body surface area includes both height and weight). The average diameter is assigned a Z score of 0. Positive Z scores reflect larger diameters, while negative Z scores reflect smaller diameters.

Most individuals (~95%) have coronary artery Z scores between -2 and +2, and are considered to have normal coronary arteries (see figure below). A coronary artery Z score **between +2.0 and less than +2.5** (i.e., 2 to less than 2.5 standard deviations above the average normalized for body surface area) is considered **dilated**. A coronary artery with a Z score **between +2.5 and less than +5.0** is considered a **small aneurysm**. A Z score **between +5.0 and less than +10.0** is considered a **large aneurysm**. A Z score of **+10.0 or greater** is considered a **giant aneurysm**.





WHY DO WE USE Z SCORES WHEN MEASURING CORONARY ARTERY INTERNAL DIMENSIONS?

Coronary artery aneurysms can be measured in millimeters (mm). However, this measurement does not account for body size. How big the coronary artery should be depends on the size of the child. For instance, a 4-mm aneurysm in a 1 year-old patient carries more severe long-term cardiovascular risks compared to a 4-mm aneurysm in a 10 year-old patient. In contrast, Z scores normalized for body surface area account for body size. This allows us to accurately assess and track aneurysm size over time (i.e., as the patient grows).