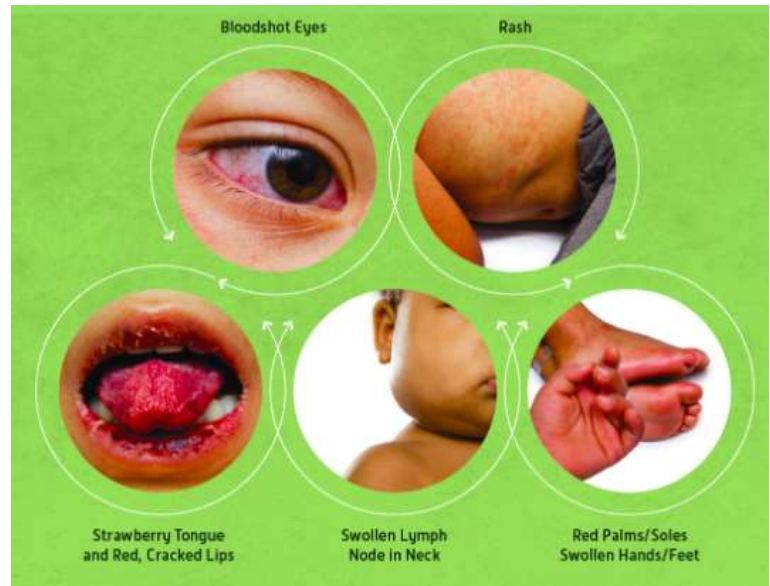


Kawasaki Disease: General Information

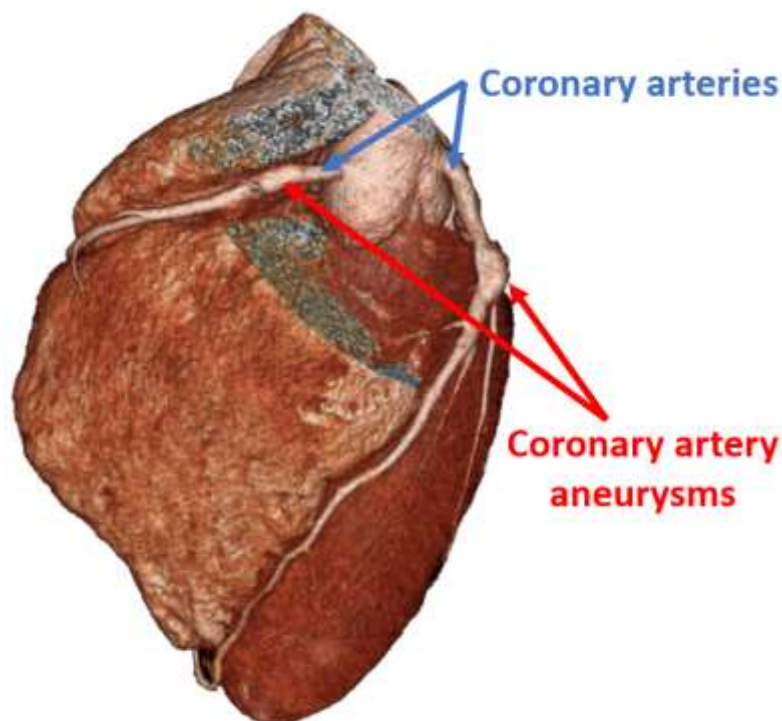
What is Kawasaki disease?

Kawasaki disease (KD) is an inflammatory disorder that affects the heart and the coronary arteries (blood vessels that supply the heart) and heart muscle. It primarily affects children. In addition to fever, KD can present with a combination of the following:

- Bloodshot eyes
- Rash
- Swollen lymph nodes in the neck
- Red, cracked lips and red tongue
- Swelling of the hands and feet



Up to 25% of untreated patients will develop coronary artery aneurysms. The inflammation from KD weakens coronary artery vessel walls, which can lead to aneurysms (ballooning) of the arteries. These coronary artery aneurysms are concerning because they can be sites for blood clot formation, and thus can increase the risk of heart attacks.



CT scan of the heart: KD patient with two coronary aneurysms.

What causes Kawasaki disease? Is Kawasaki disease hereditary?

The exact cause of KD is currently unclear. Most experts believe that KD occurs when genetically susceptible children are exposed to certain environmental triggers. Your future offspring have a greater risk of developing KD, since the same genes that made you susceptible to KD may also be passed on to them. There is no evidence to suggest that KD is contagious.

How is Kawasaki disease initially treated?

KD is initially treated with aspirin and intravenous immunoglobulin (IVIG). These help to reduce both the inflammation and the risk of developing coronary artery aneurysms.

What are the major long-term complications of Kawasaki disease?

Most people with KD do not experience any further problems with their heart or blood vessels. However, those with coronary artery aneurysms have a higher risk of heart attacks, as well as scarring of the heart and blood vessels. These individuals require life-long care by a cardiologist and may require further medical or surgical treatment.

Kawasaki Disease: Medications

Antiplatelet Drugs

Prevents platelets (cells that help blood clot) from sticking together and forming blood clots in the arteries.

Possible side effects: Bleeding, bruising

Examples:

Low-dose aspirin

Clopidogrel (Plavix)

Anticoagulation Drugs

Reduces blood clot formation in the arteries by inhibiting clotting proteins in the blood.

Possible side effects: Bleeding, bruising

Examples:

Apixaban (Eliquis)

Warfarin (Coumadin)

Enoxaparin (Lovenox)

If you take any of these medications, you should wear a medical alert and register your medical information with one of these companies in case of an emergency (such as a car accident).

Statins

Helps heal the lining of blood vessels in the heart and may prevent build-up of scar tissue in the damaged artery

Possible side effects: Muscle pain, liver inflammation

Examples:

Atorvastatin (Lipitor)

Rosuvastatin (Crestor)

Simvastatin (Zocor)

Getting Your Medications

Your prescription medications can be picked up at your pharmacy. Each prescription has a certain number of refills. If you need refills on your prescriptions, you may contact your pharmacy. You should contact your pharmacy for a refill when you have fewer than two weeks' worth of medications left.

If you have no more refills left on a prescription, then you will need to contact your doctor.

My pharmacy is: _____

Phone number: _____

My doctor is: _____

Phone number: _____

Taking Your Medications

It is *extremely important* that you take your medications at the right dose, at the right time, every time. Incorporating medications into your daily routine will help ensure that you are consistently taking your medications.

Recommendations:

- Use a weekly pill organizer
- Keep your medications/pill organizer next to something you use everyday (e.g., toothbrush, coffee maker, computer, phone charger)
- Keep some extra medications in your bag/purse at all times
- Set a daily reminder on your phone
- Use a free phone app (e.g., Medisafe) to keep track of when you take your medications



7-day pill organizers

Who Do I See Next?

Dr. John Gordon is a cardiologist at the San Diego Cardiac Center who specializes in the care and treatment of adults with coronary artery disease. He also follows many adults in his practice who had KD as children. As a member of the UCSD Kawasaki Disease Research Center, he is a co-investigator for the Adult KD Study.

Address:

San Diego Cardiac Center
3131 Berger Avenue, Suite 200
San Diego, CA 92123

Tel: (858) 244-6800

Fax: (858) 244-6809

Website: <http://sdcardiac.com/cardiac>



John Gordon, M.D.

Dr. Lori Daniels is a cardiologist who serves as the Director of the Coronary Care Unit at the UCSD Sulpizio Cardiovascular Center. She manages and treats patients who have a variety of cardiovascular issues, including adults who had KD as children. As a member of the UCSD Kawasaki Disease Research Center, she is the principal investigator for the Adult KD Study.

Address:

Sulpizio Cardiovascular Center
9434 Medical Center Drive
La Jolla, CA 92037

New Patient Registration: (800) 926-8273

Direct Clinic Line: (858) 657-8530



Lori Daniels, M.D., M.A.S.

When should my first appointment be?

Your first appointment should be *within 3 months* of your last visit with the Rady Children's Hospital KD clinic.

My first appointment is on

Date: _____

Time: _____

What should my doctor know?

Whenever you see a new doctor, you should let him/her know that you were diagnosed with KD. Provide the following details:

- When were you diagnosed with KD and how old were you?
- How and where were you treated for KD?
- What was your Z_{\max} (a measure of your maximum coronary artery internal diameter)?
- What medications, if any, do you take for your heart?
- What is the name and location of your pharmacy?
- What allergies do you have?
- When was your last echocardiogram and what were the results?

Chest Pain

Kawasaki disease patients with coronary aneurysms are at an increased risk of developing cardiovascular complications, including a heart attack (myocardial infarction). There are different kinds of chest pain, and sometimes it can be hard to distinguish between all of them. Listed below are some of the most common types of chest pain:

Heartburn/Acid Reflux (GERD)

Symptoms: Burning pain in the chest and/or throat
Can occur after eating a large meal or spicy/acidic foods
Can occur before meal time when you are hungry and the stomach is secreting acid
May experience acidic taste in the mouth, especially after lying down.

What to do: Take an antacid. Tums™ or similar tablet is safe and effective. The pain should go away within minutes after chewing and swallowing several tablets.

Hyperventilation/Panic attack

Symptoms: Heavy sensation in chest, sharp pain in chest
Feeling of breathlessness and anxiety

What to do: Slow your breathing rate either by taking in a deep breath and holding it while you count to 10 slowly or breathe in and out into a paper bag held over your mouth and nose

Costochondritis

Symptoms: Pain or tenderness when pressing down on the ribs or when taking a deep breath

What to do: Take pain relief/anti-inflammation medication (e.g. ibuprofen, naproxen (NSAIDs))

Pleuritis

Symptoms: Sharp, stabbing pain of the chest that worsens with deep breathing, especially during or after an upper respiratory infection

What to do: Take pain relief/anti-inflammation medication (e.g. ibuprofen, naproxen (NSAIDs)) and see your general medical doctor

Heart Attack (Myocardial Infarction)

Symptoms: Heavy, crushing sensation of the chest
Pain radiating up to the neck and/or down the shoulder and arm
Nausea, feeling faint

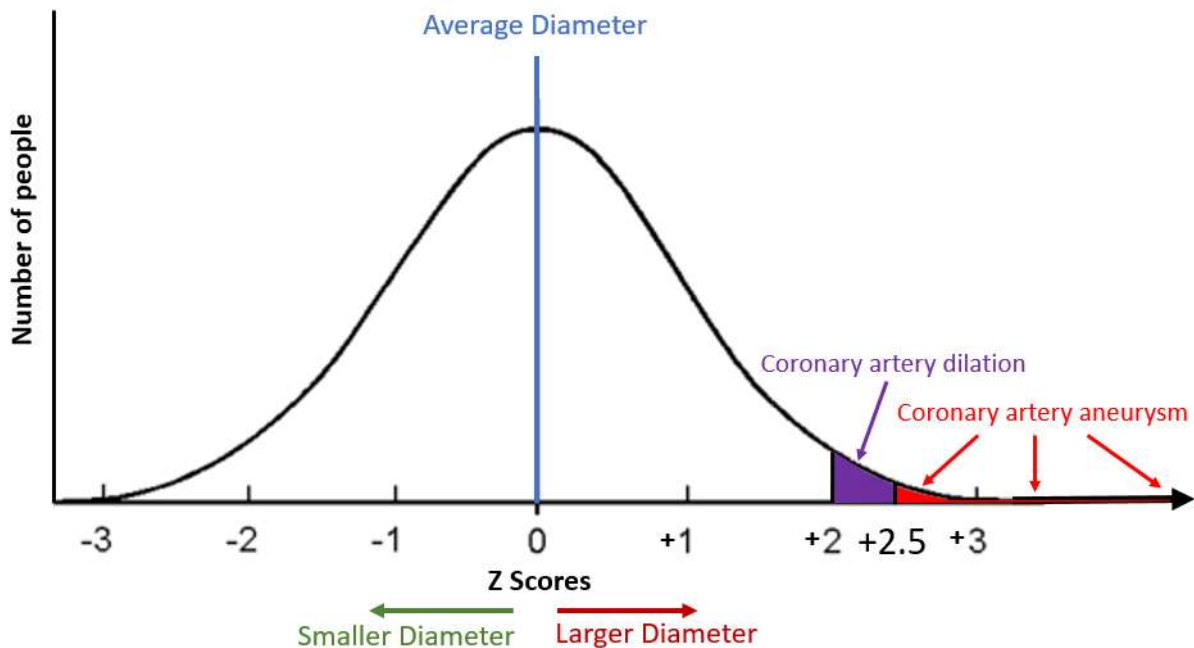
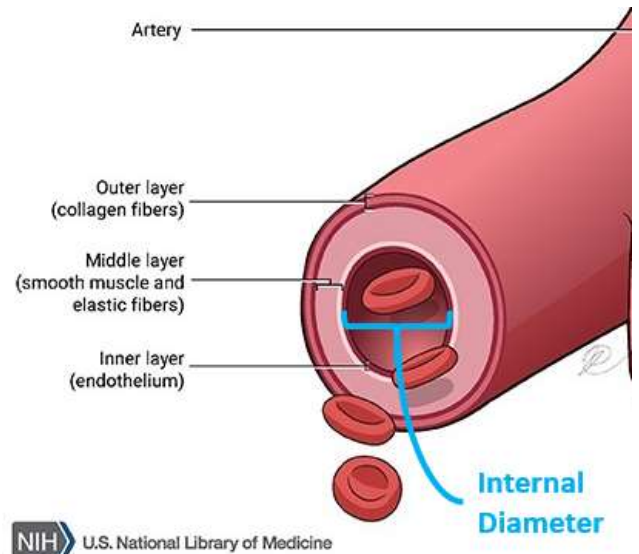
What to Do: Call 911. Take 81 mg of aspirin.

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).