Recommendations for managing Kawasaki disease (KD)

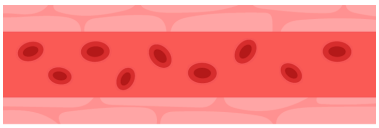
These are recommendations written by a group of doctors and patients based on research studies for people with Kawasaki disease (KD).

These recommendations suggest the best treatment for most people, but your child's situation and treatment may be different. Talk to your doctor about what treatment is best for your child.

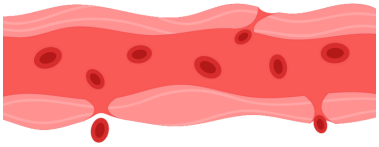
What is Kawasaki disease (KD)?

Kawasaki disease (KD): A type of vasculitis that causes inflamed blood vessels typically in children. It causes a fever that usually lasts 5 days or longer, and some or all of these symptoms:

- Red eyes
- Cracked lips and a red, swollen, bumpy tongue
- Red or swollen hands and feet
- Rash
- Swollen lymph nodes in the neck



Healthy blood vessel



Inflamed blood vessel

Words to know about vasculitis

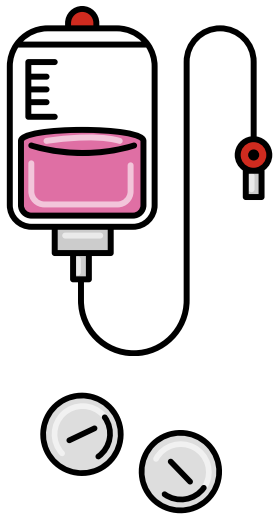
- **Vasculitis:** A group of conditions that involves inflammation (swelling) of your blood vessels.
- **Incomplete KD:** When a child has a fever for 5 or more days but less than 4 of the other symptoms.
- **Acute KD:** The first phase of KD, in which a child has a fever.
- **High-Risk KD:** When tests show that a child has a higher chance of a coronary artery aneurysm.

See a glossary of health terms at the end of this document.

Treatment recommendations

In general:

- Use IVIG as the first treatment.
- Use aspirin during acute KD, because aspirin can lower inflammation and help prevent blood clots.
- Monitor children for fevers every day for 1-2 weeks after their fever goes away. A new fever may mean KD came back.
- For children who have high-risk KD, consult a rheumatologist or KD expert before using prednisone or other immunosuppressants.



For children with acute KD and a fever that lasts after receiving one IVIG treatment:

- Use a second IVIG treatment, over prednisone.

For children with acute KD and a fever that lasts after 2 IVIG treatments:

- Use immunosuppressants or prednisone.

For children with acute KD who have arthritis that lasts after IVIG treatment and who do not have coronary artery aneurysms:

- Use NSAIDs to treat arthritis.

For children with acute KD who have arthritis that lasts after IVIG treatment and who do have coronary artery aneurysms:

Do not use NSAIDs to treat arthritis.

- Children with coronary artery aneurysms will need to use aspirin long-term. NSAIDs can make aspirin not work as well, so they will need other treatments.

Children with KD may get short-term arthritis that usually lasts 1-3 weeks.

For children with acute KD who are more likely to not respond to IVIG or who have coronary artery aneurysms:

For initial treatment, use IVIG with prednisone or immunosuppressants (such as infliximab, anakinra, or cyclosporine), over IVIG alone.

- Prednisone and immunosuppressants may lower the chance of a coronary artery aneurysm.
- These treatments should only be added to IVIG for children who have coronary aneurysms, or a higher risk of coronary aneurysms (high-risk KD).
- If a doctor is unsure if a child has high-risk KD, use IVIG alone.

For children with acute KD and suspected or diagnosed macrophage activation syndrome (MAS):

Use IVIG for KD and other treatments for MAS.

- MAS may be a health problem from KD. If MAS is not treated, it could lead to organ failure or death.

If MAS is due to KD instead of a genetic disorder, use anakinra and prednisone, over cytotoxic drugs (drugs that kill cells) to treat MAS.

- Cytotoxic drugs have more potential severe side effects and may not be needed to treat MAS due to KD.

For children with incomplete KD, treat with IVIG when they are diagnosed, over delaying treatment until day 10 of fever or later:

If a child is diagnosed with incomplete KD, treat them right away – do not wait until day 10 to see if they meet the criteria for "complete" KD.

If a child with incomplete or complete KD has their fever go away before day 10, they should still get treatment.

- Waiting to give treatment can cause serious side effects, such as a coronary artery aneurysm.

Diagnostic imaging

Get an echocardiogram with coronary artery measurements for children with:

- Suspected incomplete KD and fever, as this test can help confirm if they have incomplete KD, so they can start treatment as soon as possible.
- Unexplained symptoms of shock, as this test can help find the possible cause of shock, which may be KD.
- Unexplained MAS, as this test can see if it is KD that is causing MAS.

Health terms

A

- **Anakinra:** An immunosuppressant. Lowers inflammation (swelling) in your body.

C

- **Coronary artery aneurysm:** When a part of your coronary artery widens more than usual. Coronary arteries are the major blood vessels that supply blood to your heart.

E

- **Echocardiogram:** A test that uses ultrasound to take pictures of your heart.

I

- **Immunosuppressant:** A medicine that lowers your body's immune response to stop your immune system from causing inflammation (swelling) and damaging your body.
- **IVIG (intravenous immunoglobulin):** Immunoglobulin given as an IV through a vein in the arm. Immunoglobulin is a part of your blood that has antibodies to help you fight germs and disease.

M

- **Macrophage activation syndrome (MAS):** When 2 types of white blood cells (T cells and macrophages) become overactive. Symptoms may include low amounts of blood cells, blood clots, and liver failure. It can be a serious health problem from KD.

N

- **NSAIDs:** A type of medicine that lowers pain, inflammation (swelling), fever, and prevents blood clots. NSAIDs include ibuprofen (Advil), and naproxen (Aleve).

P

- **Prednisone:** A medicine that lowers inflammation (swelling) in your body.

S

- **Shock:** A serious condition that happens when your body doesn't get enough blood flow.

*Gorelik M, Chung SA, Ardalan K, Binstadt BA, Friedman K, Hayward K, Imundo LF, Lapidus SK, Kim S, Son MB, Sule S, Tremoulet AH, Van Mater H, Yildirim-Toruner C, Langford CA, Maz M, Abril A, Guyatt G, Archer AM, Conn DL, Full KA, Grayson PC, Ibarra MF, Merkel PA, Rhee RL, Seo P, Stone JH, Sundel RP, Vitobaldi OI, Warner A, Byram K, Dua AB, Husainat N, James KE, Kalot M, Lin YC, Springer JM, Turgunbaev M, Villa-Forte A, Turner AS, Mustafa RA. 2021 American College of Rheumatology/Vasculitis Foundation Guideline for the Management of Kawasaki Disease. Arthritis Rheumatol. 2022 Apr;74(4):586-596. doi: 10.1002/art.42041. Epub 2022 Mar 7. PMID: 35257501.

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