Top Ten Things to Know
Diagnosis, Treatment, and Long-Term Management of Kawasaki Disease

1. Kawasaki disease (KD) is an acute vasculitis of childhood that leads to coronary artery aneurysms in ≈25% of untreated cases. It accounts for 5% of acute coronary syndromes (ACS) in adults <40 years of age and is the leading cause of acquired heart disease in children in developed countries.

2. First described in Japan, KD has now been diagnosed worldwide, but the disease remains markedly, i.e. ~10 fold, more common in children in Japan.

3. KD is an acute, self-limited febrile illness that predominantly affects children <5 years of age; the incidence in boys is about 1.5 times higher than in girls.

4. The diagnosis of KD rests on the identification of principal clinical findings and the exclusion of other clinically similar entities with known causes.

5. The cause remains unknown. We have no means to prevent KD. Until these are defined, an exact diagnostic test remains elusive, and acute treatment remains somewhat empirical.

6. Careful early management of evolving coronary artery abnormalities is essential, and an increased frequency of assessments and escalation of thromboprophylaxis is necessary.

7. Although intravenous immune globulin (IVIG) is the mainstay of initial treatment, the role for additional primary therapy in selected patients is discussed. Approximately 10% to 20% of patients do not respond to initial intravenous immune globulin (IVIG Resistance).

8. The current scientific statement incorporates
   • new evidence regarding underlying pathological processes
   • an algorithm to ensure capture of incomplete KD during the effective window of therapy
   • improved management of the acute illness that includes the use of additional therapies for IVIG-refractory patients
   • greater use of Z scores for classifying coronary artery involvement
   • greater specification of long-term management based on both initial and current coronary artery involvement
   • acknowledgement of the care needs of a growing population of adults with a previous history of KD and coronary artery aneurysms

9. Effective care strategies to address psychosocial concerns and to ensure transition to uninterrupted expert adult cardiology care are essential to optimize quality of life across the lifespan.

10. Although this scientific statement provides updated and best evidence-based guidance to healthcare providers who diagnose and manage Kawasaki disease, clinical decision making should be individualized to specific patient circumstances.