1.17 KAWASAKI DISEASE

Introduction

Kawasaki Disease (KD), also known as mucocutaneous lymph node syndrome, is a multisystem inflammatory disease of childhood. It most commonly presents in children ages 6 months to 5 years but can be seen as early as 1 month of age and into early adulthood. Pediatric hospitalists must have a high index of suspicion for KD; diagnosis can be difficult, as the classic signs and symptoms may not all manifest and the presentation may mimic other febrile illnesses. Consideration for KD must be particularly high in children being evaluated for fever of unknown origin, as this is a population in which KD is commonly missed or diagnosed late. Although many organs may be affected, involvement of cardiac structures is the most concerning, often leading to persistent morbidity. Coronary aneurysms occur in up to 25% of untreated children with KD. If diagnosed and treated within the first 10 days of illness, the cardiac complications can be reduced. Therefore, it is important that pediatric hospitalists have a complete understanding of the diagnosis and treatment of KD, as well as the associated complications.

Knowledge

Pediatric hospitalists should be able to:

- Describe the range of clinical presentations for KD, including typical, incomplete, isolated cervical lymphadenopathy KD, and KD shock syndrome.
- Discuss currently established criteria and guidelines for diagnosis and treatment.
- Define incomplete KD and note age groups in which this is more common.
- Identify patients at the highest risk for coronary artery aneurysms.
- List the broad categories of alternate diagnoses and give examples from each.
- Discuss the appropriate laboratory and imaging studies that aid in diagnosis.
- Explain the pathophysiology of the disease, including the current understanding of development and manifestation of cardiac complications.
- Define refractory KD and list factors that indicate the need for further treatment.
- Compare and contrast the risks, benefits, and side effects of available treatments such as immunoglobulin, steroids, anti-platelet medications, and immunomodulators.
- Discuss the immediate and long-term follow up care needed, and the impact, if any, on physical activity and sports participation.
- List appropriate discharge criteria for KD.

Skills

Pediatric hospitalists should be able to:

- Diagnose KD by efficiently performing an accurate history and physical examination and ordering appropriate diagnostic studies.
- Consult appropriate subspecialists to assist in evaluation and treatment.
- Perform an evidence-based, cost-effective diagnostic evaluation and correctly interpret laboratory and imaging results.
- Demonstrate basic proficiency in reading electrocardiograms.
- Perform careful reassessments daily and as needed, noting changes in clinical status, and respond appropriately.
- Initiate prompt treatment once the diagnosis is established.
- Anticipate and treat complications and side effects of instituted therapies.
- Identify treatment failure (refractory KD) and institute appropriate repeat or alternate therapy.
- Coordinate care with subspecialists and the primary care provider, arranging appropriate transition and follow-up plans for after hospital discharge.

Attitudes

Pediatric hospitalists should be able to:

- Realize the importance of effective communication with patients, the family/caregivers, and other healthcare providers regarding findings and care plans.
- Realize the importance of educating patients and the family/caregivers on the natural course of the disease.
- Acknowledge the importance of collaborating with subspecialists and the primary care provider to ensure coordinated longitudinal care for children with KD.

Systems Organization and Improvement

In order to improve efficiency and quality within their organizations, pediatric hospitalists should:

- Lead, coordinate, or participate in early multidisciplinary care to promote efficient diagnosis, treatment, and discharge of patients with KD.
- Work with hospital staff and subspecialists to educate other healthcare providers regarding the diagnosis and treatment of KD.
- Lead, coordinate, or participate in community education efforts regarding KD.

References

1. McCrindle BW, Rowley AH, Newburger JW, et al. Diagnosis, treatment, and long-term management of Kawasaki Disease: A scientific statement for health professionals from the American Heart Association. *Circulation*. 2017;135:e927. https://www.ahajournals.org/doi/full/10.1161/CIR.0000000000000484?url_ver=Z39.88-2003&rfr_id=ori%3Arid%3Acrossref.org&rfr_dat=cr_pub%3Dpubmed. Accessed August 28, 2019.