

Kawasaki Disease in Two Patients with Hypoplastic Left Heart Syndrome Following Bidirectional Glenn Procedure

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Background

- Kawasaki Disease is the most common cause of acquired heart disease in the United States
- Etiology of Kawasaki Disease remains unknown but various hypotheses exist
- Cardiac involvement typically includes coronary artery disease
- Kawasaki Disease is rarely reported in patients with congenital heart disease

Case 1

- 2-year-old boy presented to the ED with one day of fever, rash and elevated inflammatory markers but was discharged home with presumed viral illness
- Re-presented at six days of illness with fever, hand edema, conjunctivitis and irritability with rising inflammatory markers
- Echocardiogram at presentation showed normal coronary arteries, normal systemic right ventricular function and no pericardial effusion
- He was treated with IVIG and aspirin with resolution of fevers
- No cardiac involvement developed over the follow-up period



Figure 1: Rash on patient in case 1 at presentation of second ER visit

Case 2

- 6-year-old boy presented with one day of fever and conjunctivitis, which was presumed viral
- Re-presented at six days of fever with associated conjunctivitis, lymphadenopathy and mucositis
- Echocardiogram showed normal systemic right ventricular function with mild dilation of the right coronary artery but no aneurysm formation
- He was treated with IVIG and aspirin with resolution of fevers
- His follow-up echocardiograms began showing resolution of coronary artery dilation around 6 months following diagnosis

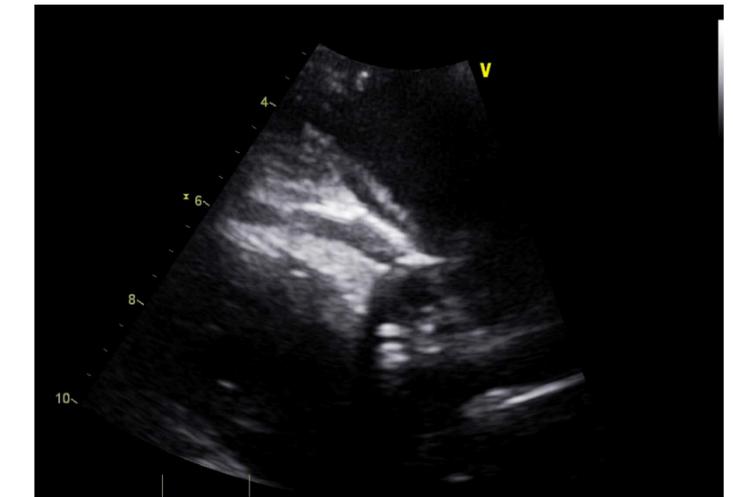


Figure 2: RCA dilation to 5.5 mm on patient in case 2

Discussion

- Only one prior report of Kawasaki Disease in a hypoplastic left heart patient
 - Following Fontan completion
- Risk for single ventricle patient theoretically having worse outcomes with Kawasaki disease
- Underlying cardiac disease is not a known risk factor for development of Kawasaki disease

Conclusion

- Kawasaki Disease has the potential for serious, life-long cardiovascular complications if not recognized and appropriately treated
- Early treatment is especially crucial in patients with complex underlying cardiac disease that are at higher risk for hemodynamic instability