



Kawasaki Disease Complicated by Coronary Artery Aneurysms: Mortality and 40-year Outcomes

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Presenter Disclosure Information



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Background

- The first patients known to have the KD are just now reaching the 4th and 5th decade of life.
- Long-term outcomes and life expectancy for children after KD remain to be determined.
- The lack of long-term prognosis data has been identified as a major source of anxiety for patients with a previous history of KD.
- This is particularly true for patients with CA aneurysms who are at risk of CV events/interventions.



Objectives

- Determine the long-term (40 years) risk of major cardiac complications after KD:
 - Revascularization
 - Symptomatic thrombosis
 - Myocardial infarction
- Estimate life expectancy up to the 4th decade of life for children with KD and compare to expected mortality.



Methods

- Inception cohort of KD patients was created:
 - Diagnosed between 1974 and 2013
 - Seen at The Hospital for Sick Children
 - Primary pediatric care centre for the Greater Toronto Area (primary catchment area)
 - Referral centre for high-risk patients
 - Majority of patients who require ongoing care after age 18 years are followed in the Peter Munk Cardiac Centre at Toronto General Hospital.
- We reviewed complete medical records for all patients at both sites in 2014.



Methods

- Freedom from all time-dependent outcomes was modelled using multiphase parametric hazard regression models.
- Life tables from Statistics Canada were used to estimate age/gender specific expected mortality for the general population.

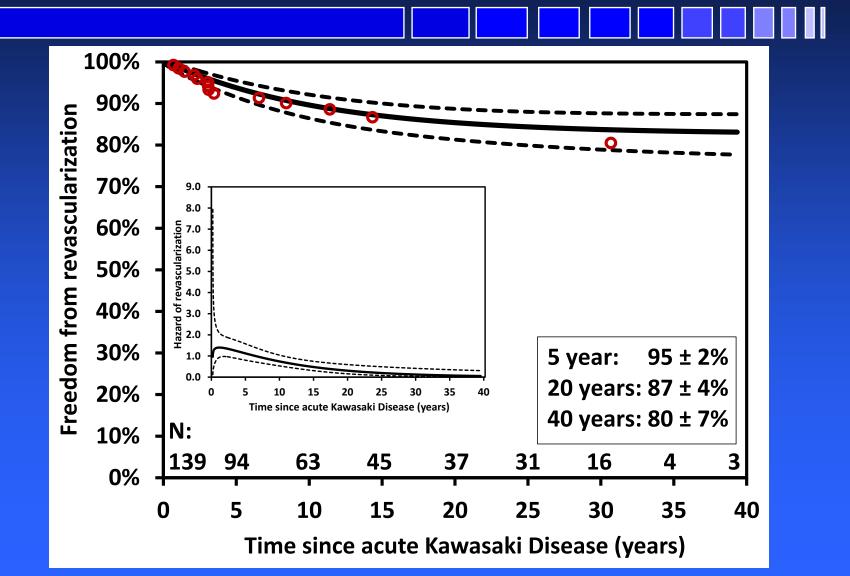


Patient Population

- 2,623 patients included in the study cohort
- 410 (16%) with CA involvement:
 - 215 CA dilatation (z > +2.5 and z < +5.0)
 - 56 medium CA aneurysms (z≥ +5.0 and z< +10.0)
 - 138 large/giant CA aneurysms (z≥ +10.0)
- Average follow-up (63% with complete adult FU):
 - 6.7 years for patients with CA aneurysms
 - 13.3 years for large/giant CA aneurysms
 - 57/34 patients with >15/25 years of follow-up
- No cardiac complications were noted in patients without large/giant CA aneurysms.

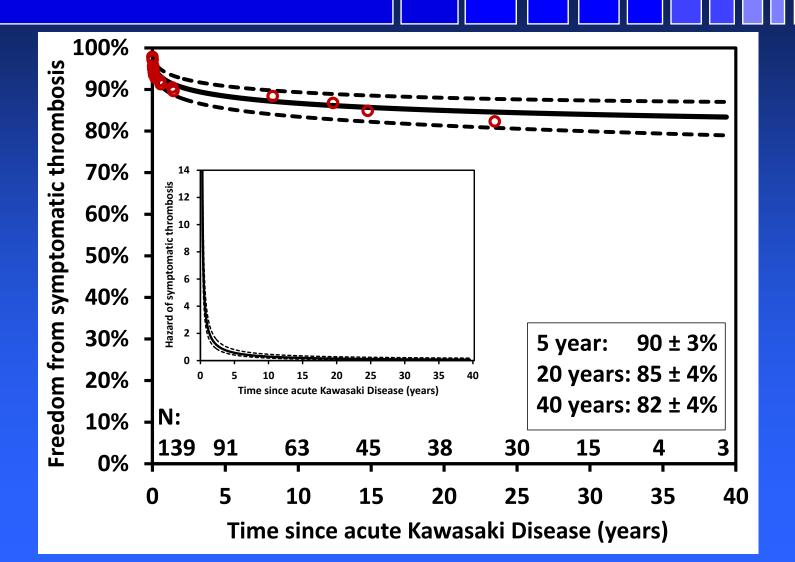
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Freedom from Revascularization in Large/Giant CA aneurysms: 14 Events



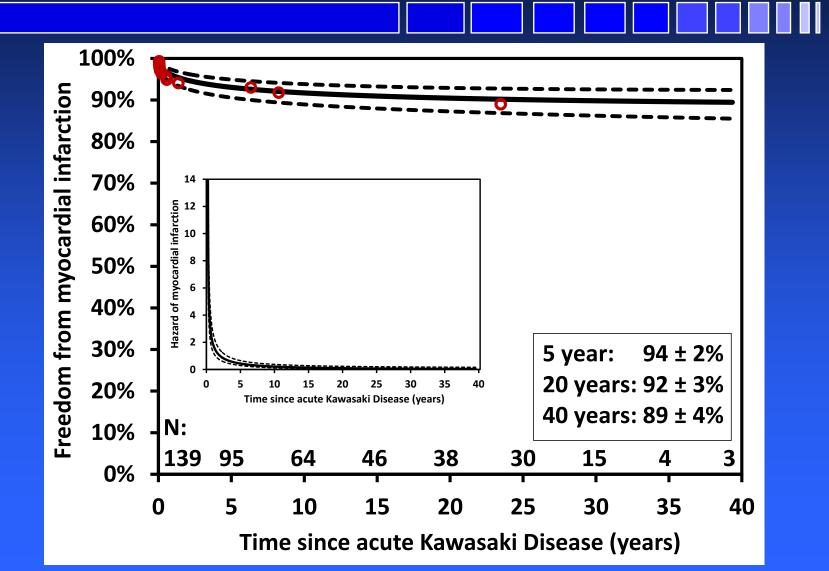


Freedom from Occlusive and/or Symptomatic CA Thrombosis in Large/Giant CA Aneurysms: 19 Events



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Freedom from Myocardial Infarction in Large/Giant CA Aneurysms: 12 Events





Death

- Patients without CA involvement:
 - 3 deaths recorded (0.1% of population)
 - 1 secondary to MAS during acute phase
 - 2 cancer-related
- No deaths recorded for patients with CA dilation or non-giant CA aneurysms.
 - Limited long-term clinical follow-up
- Patients with large/giant CAAs
 - 3 deaths recorded (2.2% of population)
 - 2 related to CAA complications (fatal MI)
 - 1 non-medical cause



Mortality Rates for Patients with Large/Giant CA Aneurysms

- At 10 years of age:
 - Mortality in KD patients 1.5%
 - Expected population mortality 0.7%
 - HR: 2.2, 95%CI: 0.3-11.5, p=0.08
- At 40 years of age
 - Mortality in KD patients: 3.1%
 - Expected population mortality 2.3%
 - HR: 1.3, 95%CI: 0.4-4.0, p=0.37



Conclusions

- KD patients without large/giant CA aneurysms are not at substantial risk of CV events or interventions.
- Risk for major cardiac complications seems to be limited in the 2nd and 3rd decades of life.
- Despite being at risk of myocardial infarction and revascularization, patients with large/giant CA aneurysms had a life expectancy similar to the general population.
- Additional follow-up will be necessary to define trends beyond the 4th decade of life.