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

C. Manlhiot, A. Crean, N. Fernandopulle, B. Lew, N. Chahal, B.W. McCrindle

Division of Cardiology, Department of Pediatrics, University of Toronto, Labatt Family Heart Centre, The Hospital for Sick Children, Toronto, Canada

Peter Munk Cardiac Centre, Toronto General Hospital, University Health Network, University of Toronto, Toronto, Canada

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C. Manlhiot, HBSc

Kawasaki disease complicated by coronary artery aneurysms: mortality and 40-year

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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.
• 2,623 patients included in the study cohort
• 410 (16%) with CA involvement:
  • 215 CA dilatation \((z > +2.5 \text{ and } z < +5.0)\)
  • 56 medium CA aneurysms \((z \geq +5.0 \text{ and } z < +10.0)\)
  • 138 large/giant CA aneurysms \((z \geq +10.0)\)

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

- N: 139, 94, 63, 45, 37, 31, 16, 4, 3
- 5 year: 95 ± 2%
- 20 years: 87 ± 4%
- 40 years: 80 ± 7%

Hazard of revascularization:

- Time since acute Kawasaki Disease (years): 0, 5, 10, 15, 20, 25, 30, 35, 40
- N: 139, 94, 63, 45, 37, 31, 16, 4, 3
- 5 year: 95 ± 2%
- 20 years: 87 ± 4%
- 40 years: 80 ± 7%

Freedom from revascularization:

- Time since acute Kawasaki Disease (years): 0, 5, 10, 15, 20, 25, 30, 35, 40
Freedom from Occlusive and/or Symptomatic CA Thrombosis in Large/Giant CA Aneurysms: 19 Events

Hazard of symptomatic thrombosis

Time since acute Kawasaki Disease (years)

N:

0 5 10 15 20 25 30 35 40

0 2 4 6 8 10 12 14

0%
10%
20%
30%
40%
50%
60%
70%
80%
90%
100%

5 year: 90 ± 3%
20 years: 85 ± 4%
40 years: 82 ± 4%

Time since acute Kawasaki Disease (years)
Freedom from Myocardial Infarction in Large/Giant CA Aneurysms: 12 Events

Time since acute Kawasaki Disease (years)

N: 139 95 64 46 38 30 15 4 3

5 year: 94 ± 2%
20 years: 92 ± 3%
40 years: 89 ± 4%
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.