The spectrum of cardiovascular lesions requiring intervention in young adults after Kawasaki disease

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San Diego Adult KD Collaborative

154 individuals age ≥ 16 years with a history of KD enrolled
» Cohort 1: 63 participants (41%) originally diagnosed with KD and followed at our center

» Cohort 2: 91 participants (59%) referred from other centers or self-referred



• 20 subjects (13%) required cardiovascular interventions:

- » 12 diagnosed with KD in childhood,
- » 7 with history of a KD-compatible illness, and
- » 1 with proximal coronary artery aneurysms compatible with KD

9 had percutaneous interventions and 11 had surgery

- » Age at first intervention: 4 53 years
- 14/20 (70%) participants were asymptomatic until experiencing a major CV event:
 - » 8 had acute MI
 - » 3 had angina
 - » 1 presented with end-stage congestive heart failure requiring cardiac transplantation, and
 - » 2 presented with extremity claudication.

Challenges

 Aneurysms with large clot burden » Assess vessel size » Medical and mechanical therapy Dense calcification • Timing? • CABG vs PCI ? Peripheral Artery Disease



Giant aneurysms with large clot burden

Unrecognized, Untreated KD in Childhood

 32 year old Laotian male presents with two hours of chest pain and 2 mm inferior lead ST segment elevation No history of tobacco use, diabetes, hyperlipidemia, or family history of premature MI







Two months later...



Case #2 Dense Calcification

"Remodeled" aneurysm ≠ normal vessel

29 yo. Palestinian male presented with increasing symptoms of shortness of breath, weight gain, and fatigue.

PMHx: Kawasaki disease at age 2 yrs.

- 2D echo: 4-6mm bilateral, fusiform aneurysms
- Pericarditis requiring pericardiocentesis,
- Serial echoes: "resolution of aneurysms", normal function 2 years post-KD onset

Pt. lost to follow-up, competed in high school sports







Dense calcification in LAD

Ossification in arterial wall with bone marrow elements







Case #3

IVUS required for accurate assessment of vessel lumen size in the setting of MI

37 year old Vietnamese man presented with chest pain and inferior lead ST elevation on ECG

- » Parental history revealed KD-compatible illness age 6 years
- » Cardiac Risk Factors: smoking, hyperlipidemia







Six weeks later, he returns with unstable angina...





Cases #4 and #5

Symptomatic peripheral vascular stenoses

KD vascular lesions are distinct from atherosclerosis



"Lotus root" recanalization with calcification













Enlarging LAD aneurysm

Enlarging Giant Aneurysm

Acute KD age 6 years

- Complicated by 8mm RCA aneurysm and 9 mm LMCA and LAD aneurysm
- Treated late with IVIG, subsequently managed with warfarin + ASA
- Asymptomatic
- At age 16 years: serial angiograms confirmed that the LAD aneurysm was enlarging



Aneurysm Trimming

Surgery for 30 mm LAD aneurysm:

- » LIMA to LAD
- Pericardial patch directing the LM into the Circ
- » RIMA to RCA



Progressive Stenosis

Progressive Coronary Stenosis

Acute KD at age 18 months

- 8 mm LAD aneurysm, normal RCA
- Maintained on warfarin and ASA
- Age 16 years, CTA showed stenosis of inlet and outlet of the LAD aneurysm
- Serial CTA's demonstrated progressive narrowing



Clinical Challenge

Is this a significant stenosis?If so, how should it be treated?

Playback Time	
2:03	
FFR	0.74
Pd/Pa	0.74
Pa:iPa	73: 90
Pd:iPd	54: 72
Pa-Pd(p)	14
HR	86
List of Runs	FFR
10:08:21 AM	0.74



Conclusions

 Large thrombus burden can be managed with combination PCI and pharmacologic therapy

 Even small to moderate-sized aneurysms that "normalize" by echocardiography in childhood can lead to stenosis and thrombosis decades later.

Conclusions

- Failure to assess the extent of calcification may lead to suboptimal procedural outcomes.
- Coronary interventions without intravascular ultrasound may result in underestimation of vessel lumen diameter and under sizing stents.
- Patients with symptomatic peripheral aneurysms may benefit from endarterectomy or resection.
- Clinical issues are complex! Further studies are needed to guide best clinical practice.

