Cardiac MR Detection of coronary artery lesion in Children with Kawasaki Disease

XIHONG HU

RADIOLOGY DEPARTMENT

CHILDREN'S HOSPITAL,FUDAN UNIVERSITY

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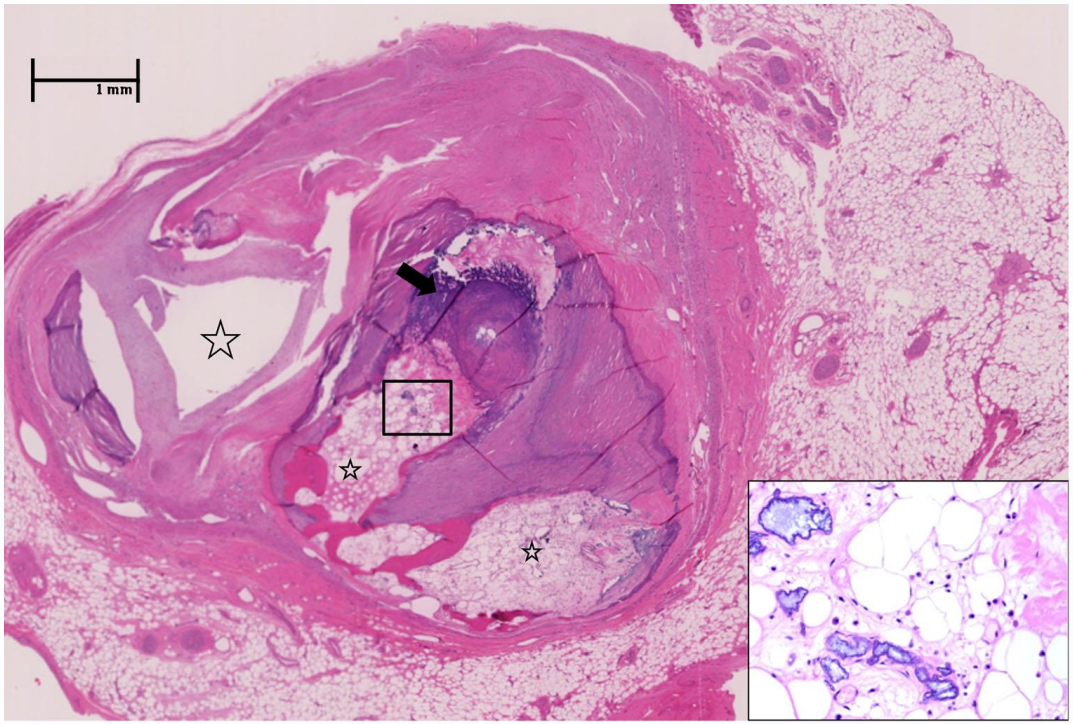
- Kawasaki disease (KD) is an acute paediatric vasculitis that mainly affects children younger than 5 years old
- Exact cause is unknown, it is thought to be caused by an infectious agent in genetically predisposed children
- This fits with the observed epidemiology of seasonal occurrence throughout the northern hemisphere, the increased incidence in children of Asian descent
- In about 25% of untreated patients, coronary artery aneurysms will develop during the acute phase of KD. Using high-dose intravenous immunoglobulin as effective treatment, the percentage of coronary artery aneurysms has dropped to 5–7% for patients treated within 10 days after fever onset

- Coronary artery aneurysms formation is an influx of inflammatory cells, leading to dissociation and disruption of the medial and internal elastic lamina layer
- Lumen of the artery returns to its normal size, the artery wall remains damaged, although the extent of the damage varies among patients
- KD-related vasculopathy during follow-up with characteristic myointimal proliferation and/or layering of thrombus can result in progressive stenosis, which may lead to ischemic cardiomyopathy

- Calcification of the damaged artery is progressive after the acute phase of the disease and may develop in coronary arteries with lesions that persist or, although rarely, in transiently dilated arteries when the vessel lumen has normalized

- The extensive calcification of the coronary artery wall is typical of the pathology observed in the remodelled lesional vessel wall

- It should be emphasized that all patients who develop coronary artery aneurysms carry a lifelong increased risk for coronary thrombosis and stenotic coronary lesions that may result in myocardial ischemia, infarction, and sudden death



Coronary artery lesion in Kawasaki disease during follow-up.
Extensive calcification (arrow) with ossification and bone marrow elements (insert, 400×) in the thrombosed and re-canalized left anterior descending artery from the explanted heart of a 29-year-old man who suffered from Kawasaki disease at age 3 years.
Characteristic 'lotus root' appearance of the artery results from thrombosis with recanalization (stars)

Definitions of coronary artery abnormalities (Japanese Ministry of Health)

Small aneurysm or dilatation	Localized dilatation with ≤4 mm internal diameter in children ≥5 years, internal diameter of a segment measures <1.5 times that of an adjacent segment
Medium aneurysm	Aneurysms with an internal diameter of >4 mm to ≤8 mm in children ≥5 years, the internal diameter of a segment measures 1.5 to 4 times that of an adjacent segment
Giant aneurysm	Aneurysms with an internal diameter ≥8 mm in children ≥5 years, the internal diameter of a segment measures >4 times that of an adjacent segment

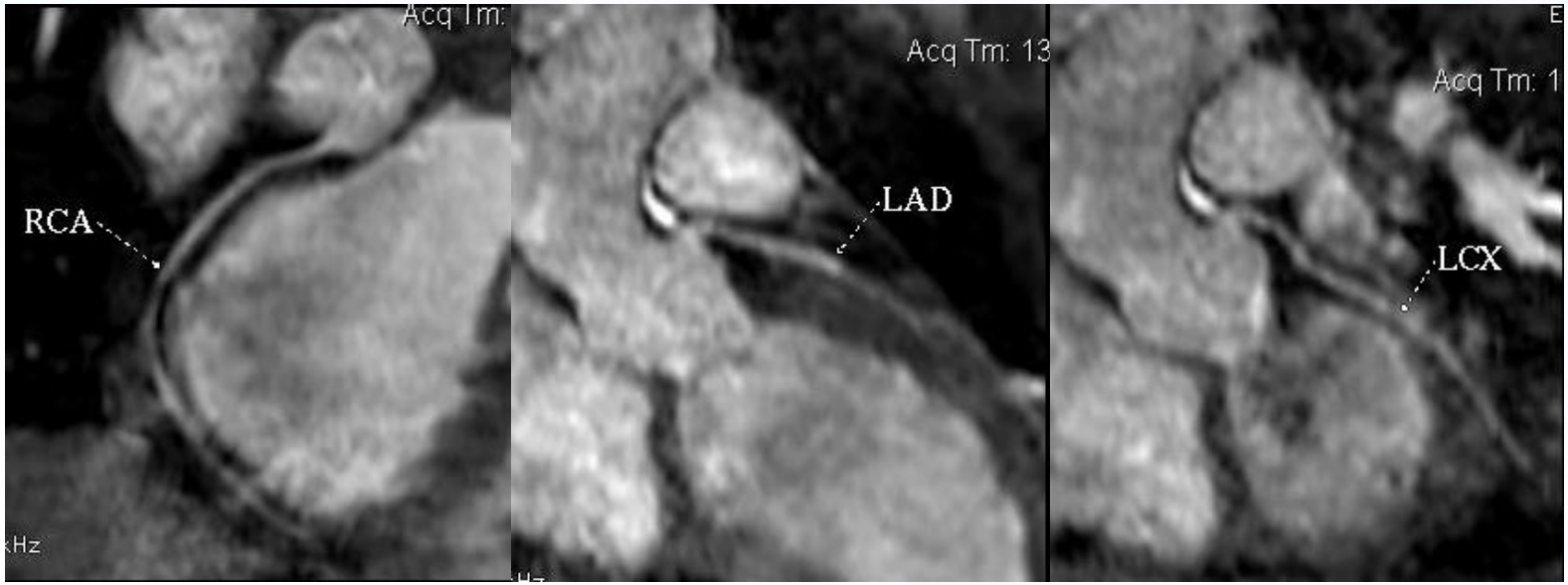
- Coronary artery imaging in children is frequently challenging due to small size, high heart rates, and motion artifacts from cardiac pulsation, respiration, which results in technical or procedural difficulties

- Imaging modalities for evaluating coronary arteries include catheter angiography, echocardiography, CT, and MRI. Recent technical advancements in CT and MRI for evaluating the coronary arteries are extremely valuable

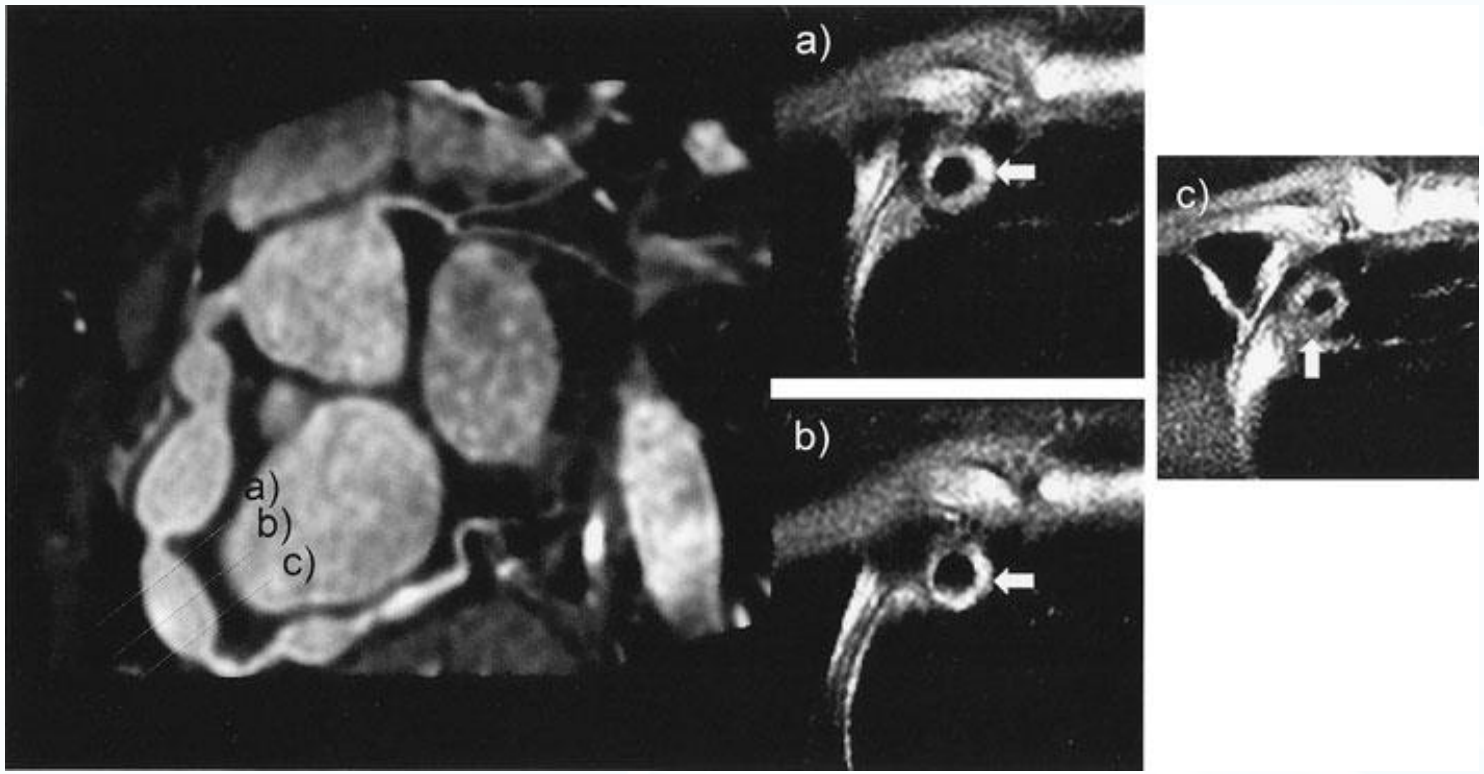
Several imaging techniques have been proposed

Imaging technique	Advantage(s)	Disadvantage(s)
Echocardiography	Non-invasive Cheap	Distal coronary arteries not visible
CAG	Complete image of coronary tree	Invasive, possible complications Need of anaesthesia Radiation exposure
CMRI	Visualization of distal aneurysms Functional assessment	Need of anaesthesia in younger children
CT-angiography	Visualization of distal aneurysms and stenosis Widely available	Radiation exposure
CT-calciumscore	Visualisation of calcifications late after disease Low-radiation exposure	Not suitable for aneurysm information Only applicable late acute disease

3D whole-heart MRA



Coronary artery lumen and aneurysm wall



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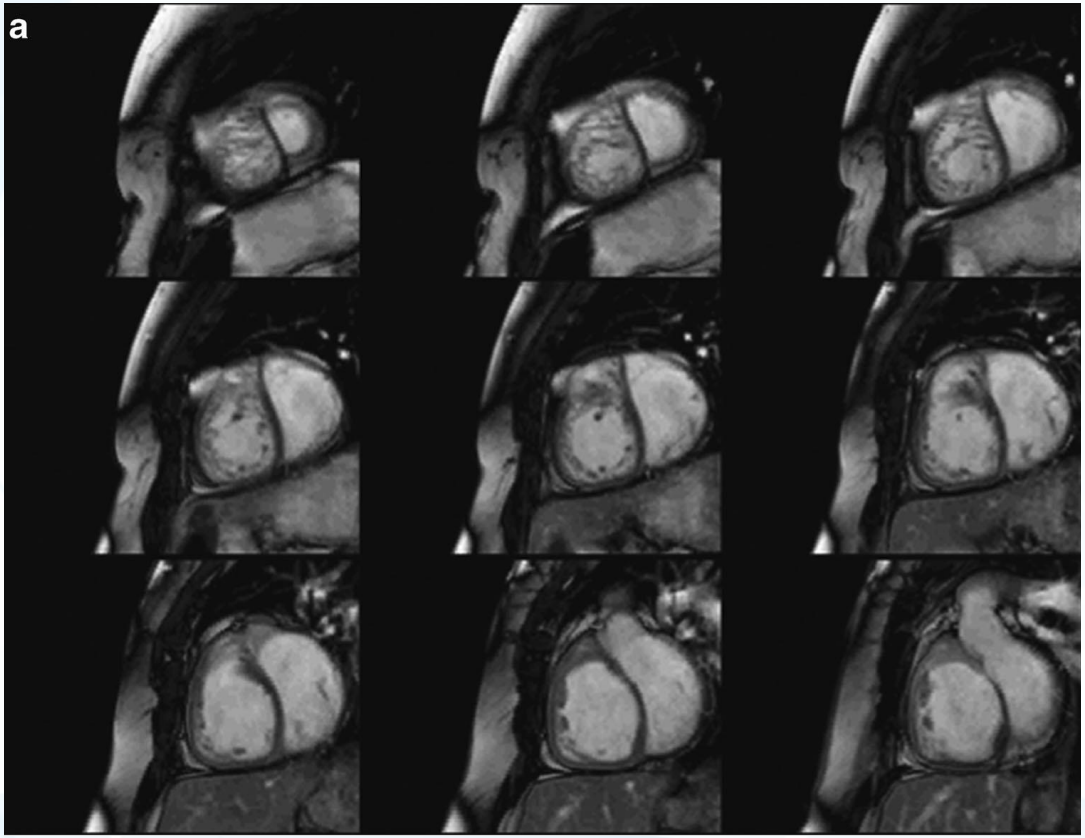
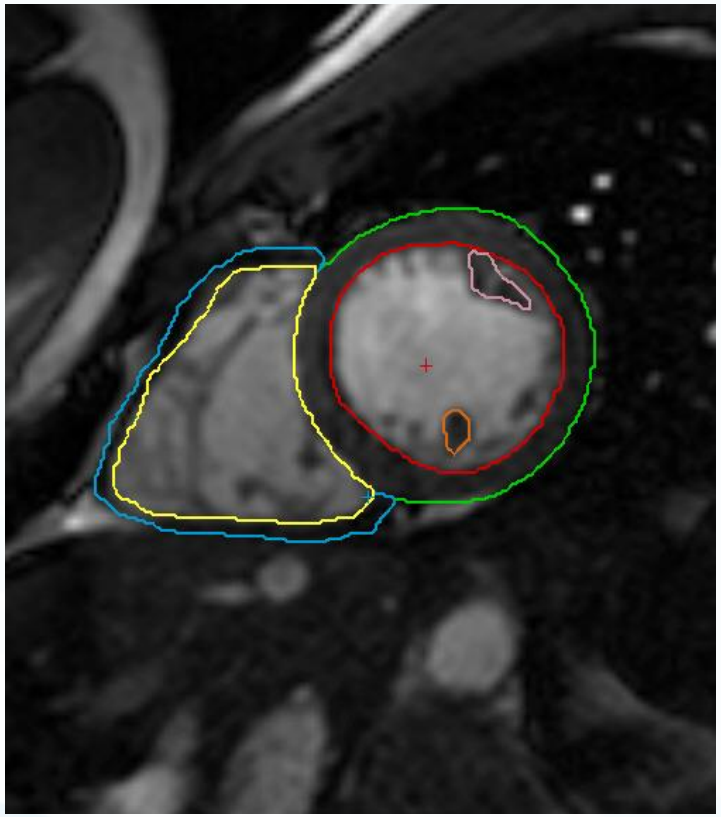
Cardiac function



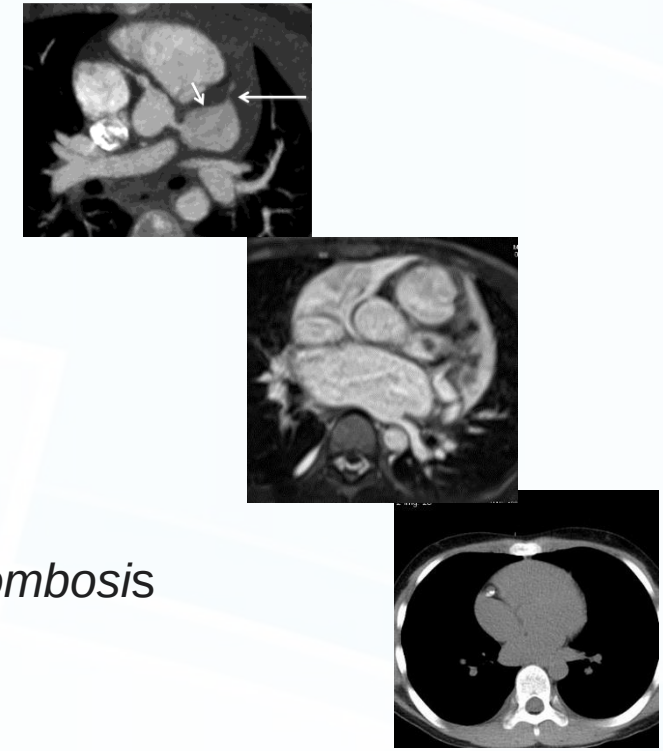


Cardiac function

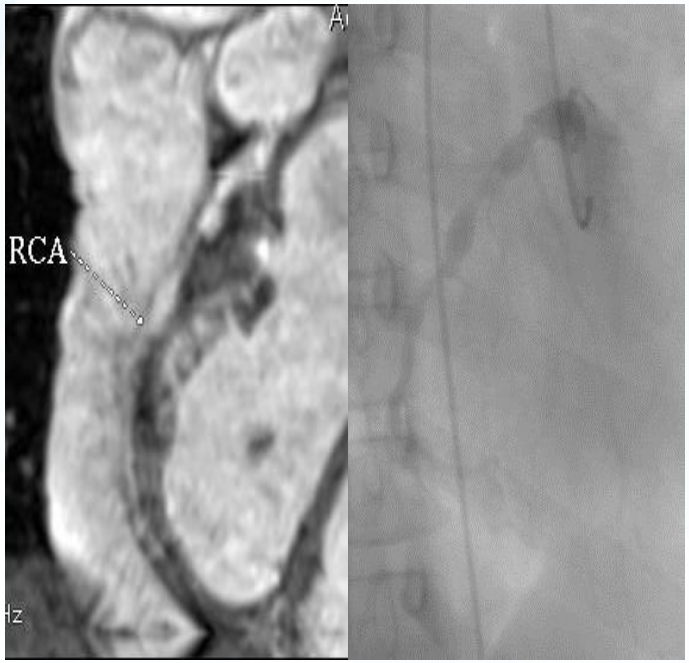
	result	normal	units
Ejection fraction EF		56-78.00	%
End Diastolic Volume EDV		52-141.00	Ml
End systolic Volume ESV		13.0-51.00	Ml
Stroke Volume SV		33-97.00	Ml
Cardiac output CO		2.65-5.98	l/min
Myocardial Mass(at ED)		75-175.00	G
Myocardial Mass(AVg)		75-175.00	g

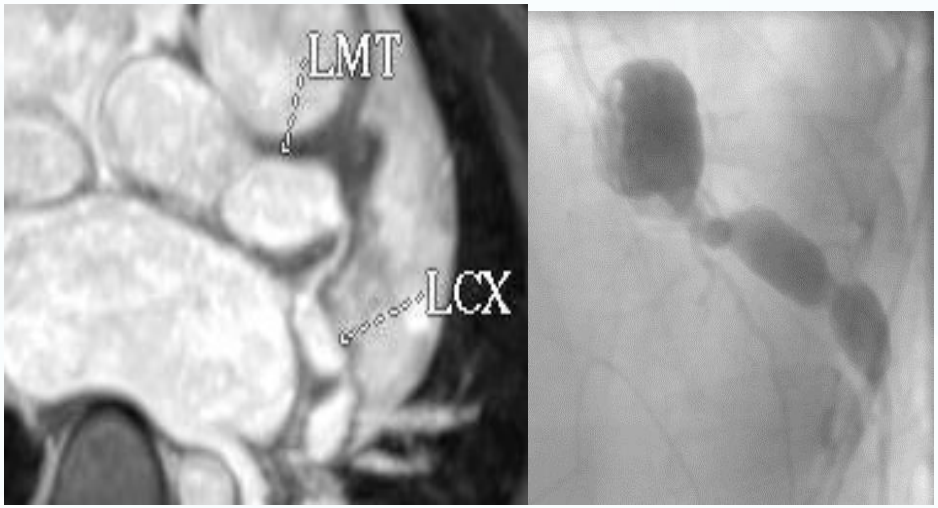
coronary artety lesion:

- *Coronary artery aneurysm*
- *Coronary artery stenosis*
- *Coronary artery calcification, thrombosis*

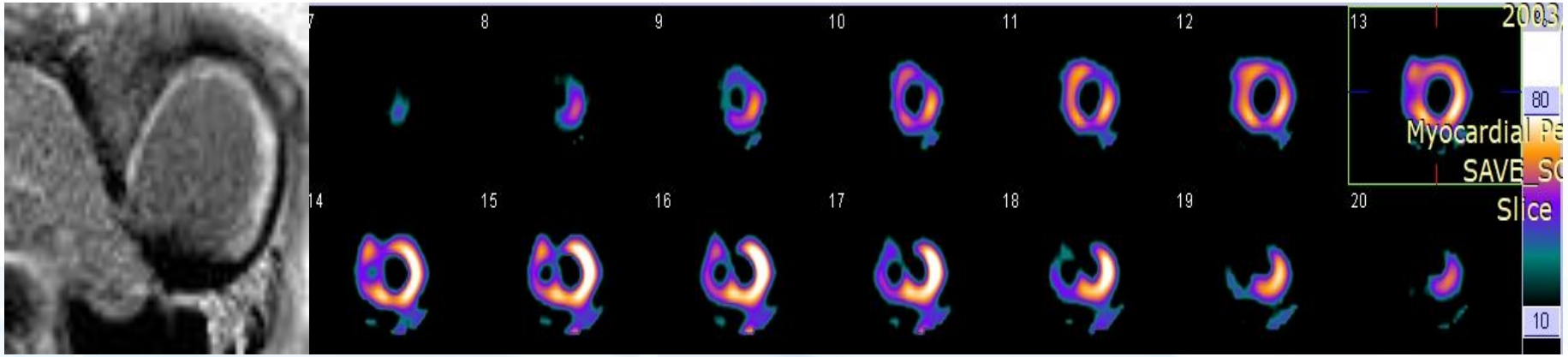


Case 1

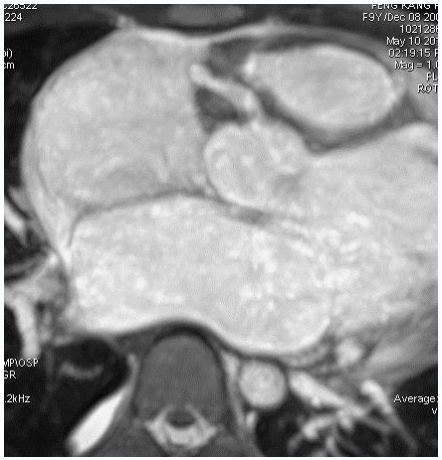


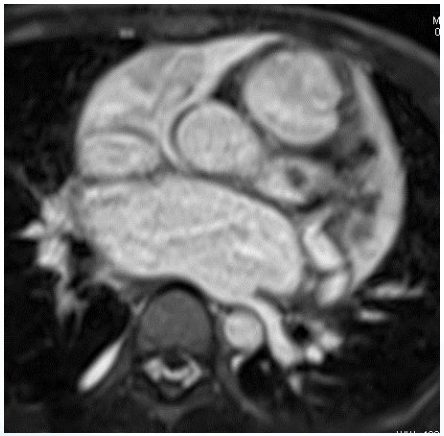


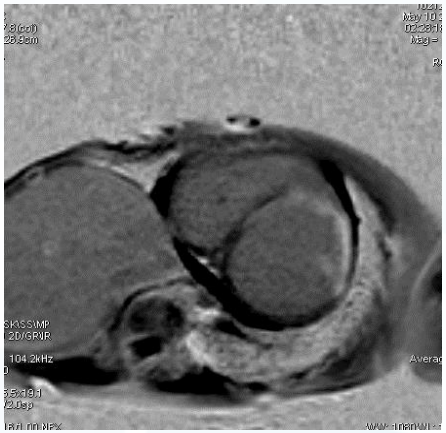
7y, girl, EF20%



Case 2





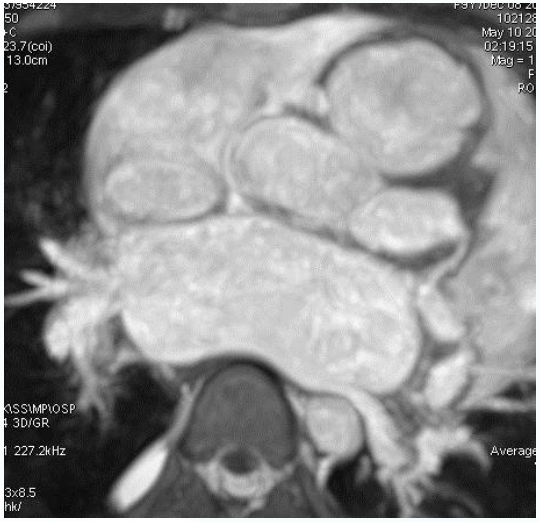


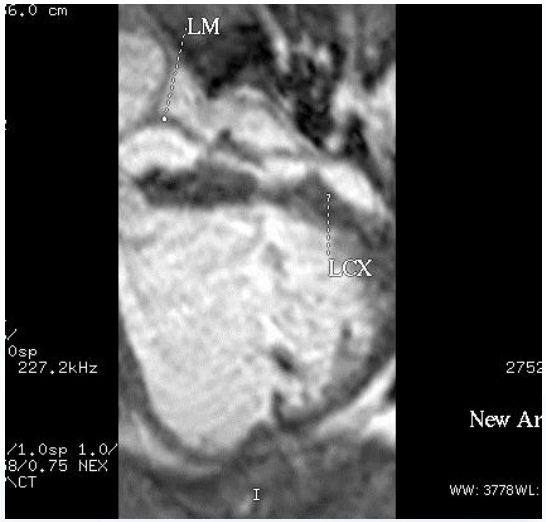
9y, girl, EF40%

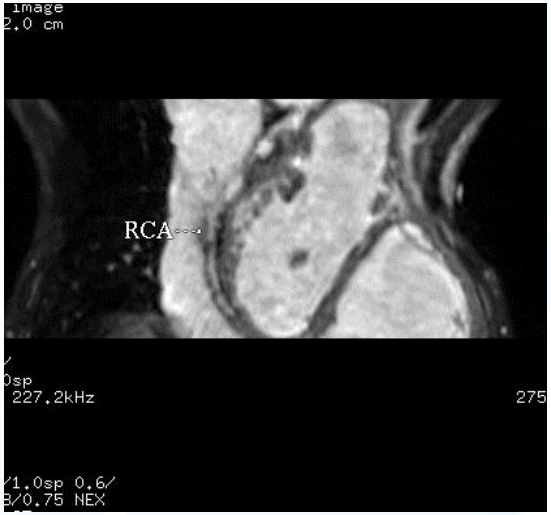
Case 3

4y, boy, EF45%

Case 4



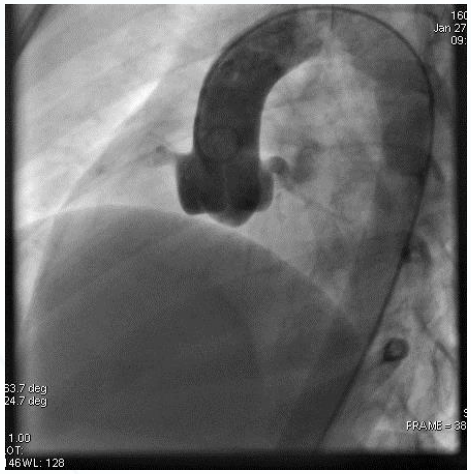
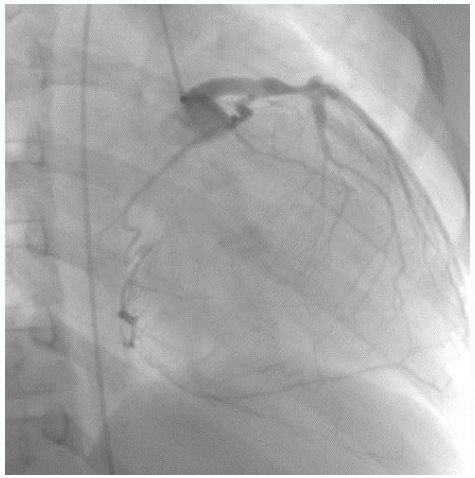
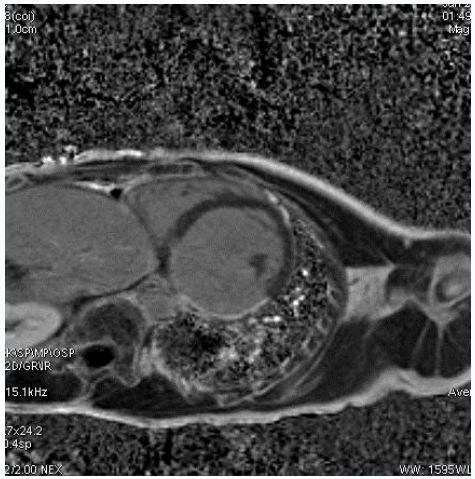




5y, boy, EF40%
coronary artery aneurysms

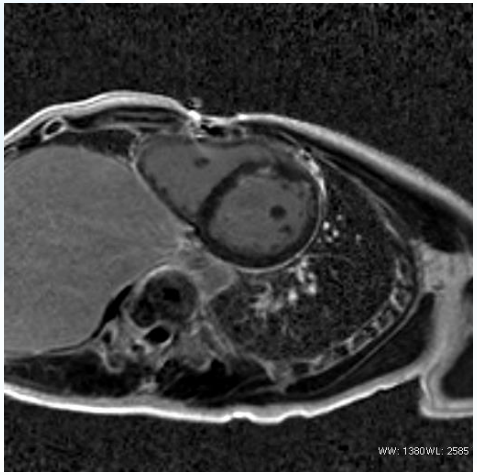
Case 5

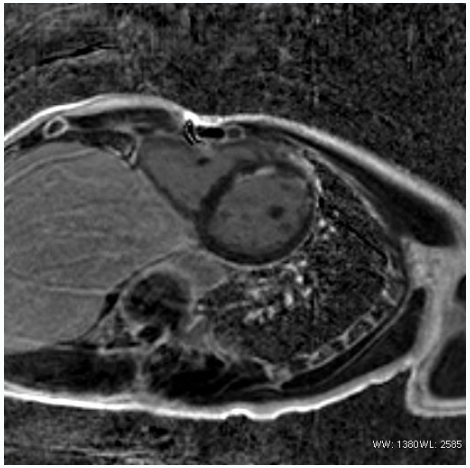


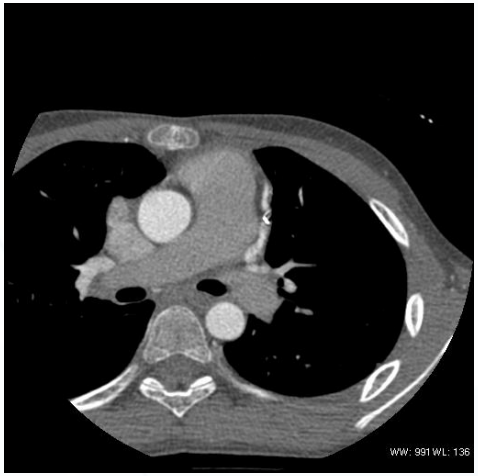





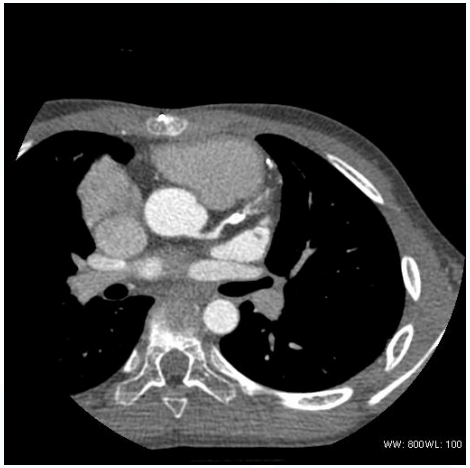
4y, boy, EF45%
coronary artery aneurysms, ischemic cardiomyopathy

Case 6









6y, boy, post-CABG

- CMRI is a robust imaging technique that is being widely adopted for the evaluation of coronary artery disease

- Coronary artery evaluation can be easily combined with the assessment of ischemia, infarction, ventricular dimensions, and function within the same procedure, for which CMRI has become the standard in children with Kawasaki disease


Thank you !