川崎病

・小孩高燒不退，請注意！

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川崎病

- 台灣現階段最常見的小兒後天性心臟病
- 1961年，日本川崎富作醫師首先發現
- 病因：不明
- 年齡：好發於3月～5歲 幼兒

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川崎病的診斷標準

- 發燒超過五天
- 眼結膜炎
- 嘴唇乾裂泛紅、草莓舌、咽喉炎
- 手腳掌初期紅腫，恢復期指（趾）端脫 皮
- 紅疹
- 頸部淋巴結腫大

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川崎病的後遺症

- 川崎病的六大症狀，即使不經治療也會隨時間逐漸消失，然而心臟的破壞卻默默地持續進行中
- 根據統計25%的病人會形成冠狀動脈血管瘤。嚴重者導致心肌缺氧、心肌梗塞或動脈血管瘤破裂而突發死亡。

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Letter to the Editor

Too big became too small

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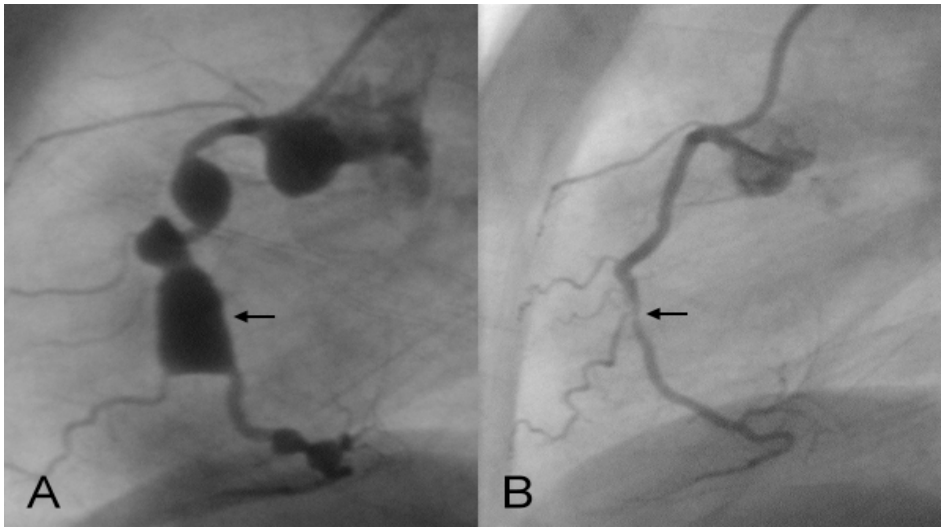
Aneurym **2 yr** → **Stenosis**

A 5-month-old boy presented with fever for 8 days in September 1999. Physical examination revealed bilateral conjunctival injection, a strawberry tongue, erythema and cracking of lips, edema of hands and feet, and diffuse erythematous skin rash. Echocardiography showed dilatations of bilateral coronary arteries. Under the diagnosis of Kawasaki disease, 2 g/kg γ-globulin was administered intravenously but the fever persisted. The 2nd course of γ-globulin was given 2 days later and the fever subsided gradually. He received oral anti-platelet treatment of aspirin and dipyridamole. The first coronary angiography was performed 3 months later and showed five aneurysms (6.8 mm, 6.1 mm, 8.8 mm, 3.4 mm and



川崎病的治療

- 免疫球蛋白：(2 g/kg iv 12 hr)
  - 於急性期使用可有效控制心臟發炎，預防冠狀動脈血管瘤的發生或改善其嚴重度
- 阿斯匹靈：
  - 高劑量 60 mg/kg/day tid or qid 可控制心臟發炎
  - 低劑量 3-5 mg/kg/day qd 可預防冠狀動脈栓塞，改善心肌血液循環

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Infection

Susceptible genes

Immune response

Vasculitis

Kawasaki disease

原因不明

診斷不清

治療明確

預後差很多

非常重要

值得研究的病

When initial treatment fails

- 10% of patients
- IVIG again 2 gm/kg
- Steroid
  - 30 mg/kg/day IVD 2-3 hours, for 1-3 consecutive days

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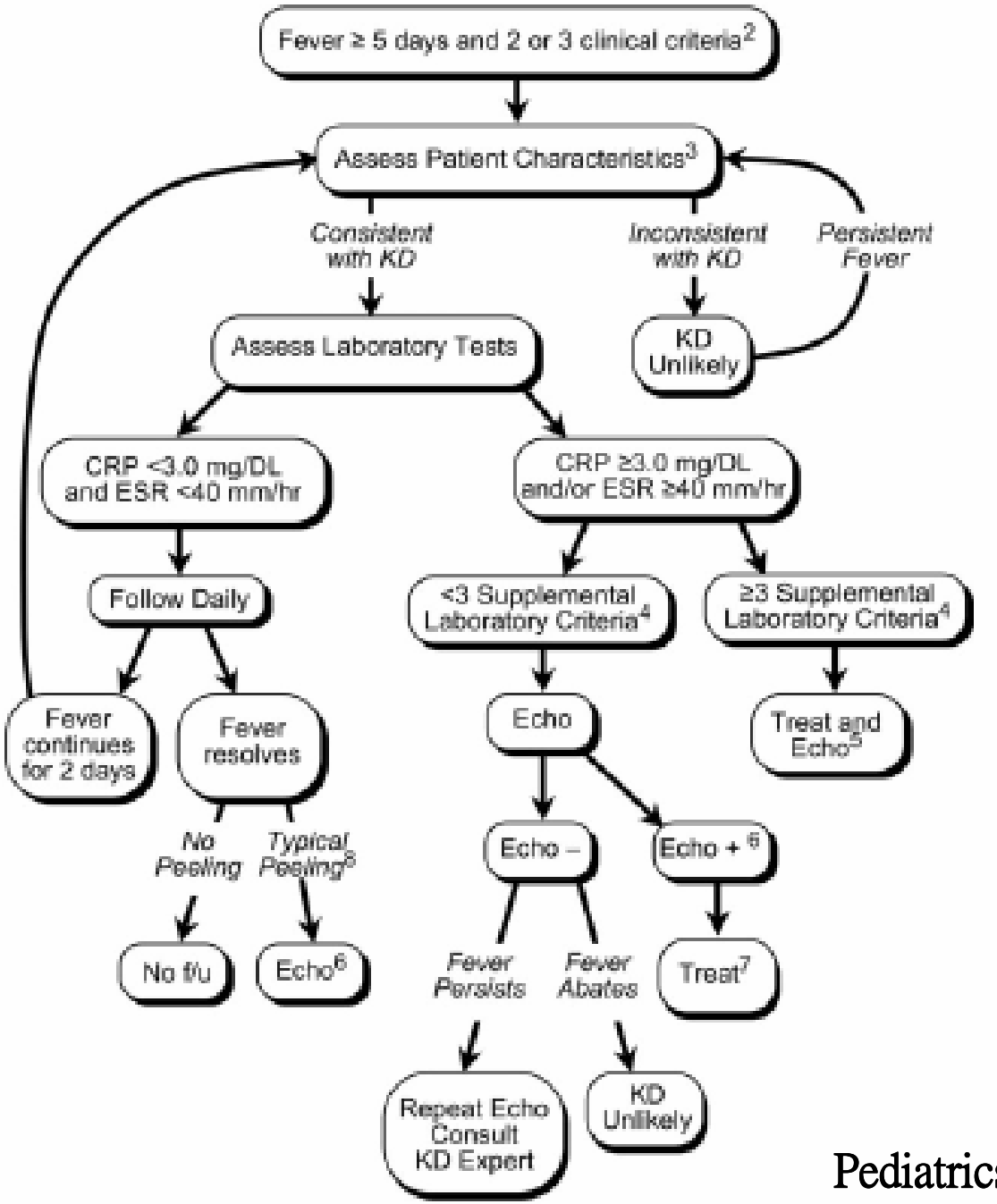
Evaluation of Suspected Incomplete Kawasaki Disease (KD)[1]



Pediatrics 2004; 114: 1708-33

Supplement Laboratory criteria

- Albumin <= 3 g/dL
- Anemia for age
- Elevation of alanine aminotransferase
- Platelet after 7 days >= 450,000 / mm^3
- Blood cell count >= 15,000 / mm^3
- Urine >= 10 white cells /HPF

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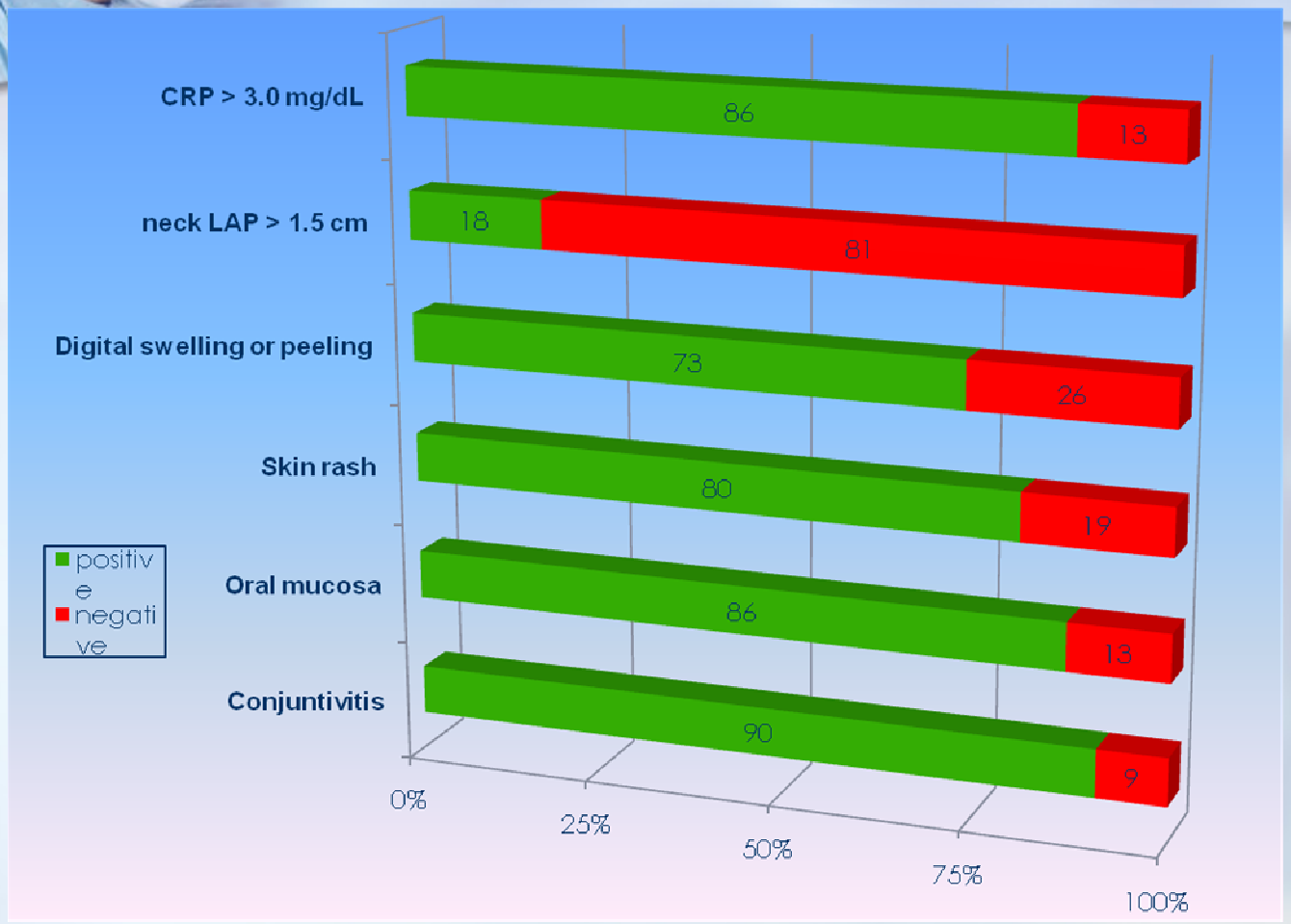

		Meeting modified criteria	
		No	Yes
Meeting criteria	No	19	21
	Yes	2	57

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Differential Diagnosis

- Viral infections
  - Measles, adenovirus, enterovirus, EBV
- Scarlet fever
- SSSS
- Toxic shock syndrome
- Bacterial cervical lymphadenopathy
- Drug allergy
- Steven-Johnson syndrome
- JRA
- Rocky Moutain spotted fever
- Leptospriosis
- Mercury hypersensitivity reaction

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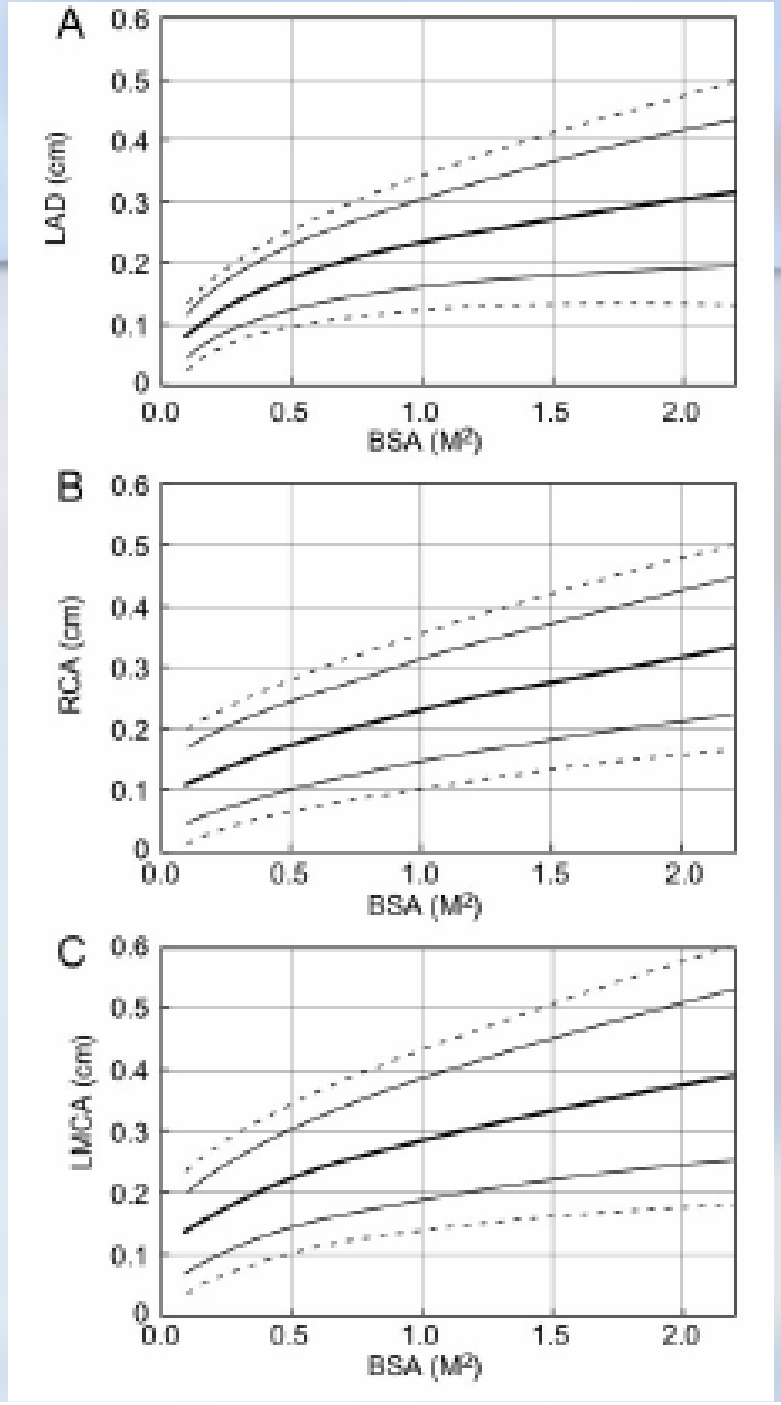

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Risk Stratifications (AHA)

Risk Level	Therapy	Physical Activity
I (no coronary artery change at any stage of illness)	Not beyond 1st 6-8 wks	No restriction beyond 1st 6-8 wks
II (transient coronary artery ectasia disappears within 1st 6-8 wks)	Not beyond 1st 6-8 wks	No restriction beyond 1st 6-8 wks
III (1 small-medium coronary artery aneurysm/major coronary artery)	Low dose aspirin (3-5 mg/kg/day), at least until aneurysm regression documented	For patients < 11 y/o, No restriction beyond 1st 6-8 wks; 11-20 y/o guided by biennial stress test; contact or high-impact sports disccouraged
IV (>= I large or giant aneurysm, or multiple or complex aneurysm in same coronary artery, without obstruction	Long term antiplatelet and warfarin (INR 2-2.5) or LMWH (antifactor Xa 0.5-1.0 U/mL) should be combined in giant aneurysms	Contact or high-impact should be avoided; physical activity guided by stress test
V (coronary artery obstruction)	Long term aspirin; warfarin or LMWH if giant aneurysm persists; Consider beta-blockers	Contact or high-impact should be avoided; physical activity guided by stress test

New Concepts from IX IKDS

- Humane genome map
- Infliximab (TNF-alpha Ab) for refractory KD
- Reconsidering of steroid in acute therapy

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