# 冠動脈障害基準化に関する小委員会英文報告

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本委員会は、昨年度に"川崎病による心血管障害の診断基準"をすでに報告したが、本年度は、この基準の英訳をおこなった。英文はLos AngelesのDr. Takahashi、ChicagoのDr. Schulmanの手をわずらわせた。記して感謝する。

# Report of Subcommittee on Standardization of Diagnostic Criteria and Reporting of Coronary Artery Lesions in Kawasaki Disease

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Research Committee on Kawasaki Diesease (Committee Chairman: T. Kawasaki) Ministry of Health and Welfare

# DIAGNOSTIC CRITERIA OF CARDIOVASCULAR LESIONS IN KAWASAKI DISEASE

The following criteria for the diagnosis of cardiovascular lesions of Kawasaki disease are meant to serve as a general guideline. In evaluating individual patients, one must use judgment on a case-by-case basis. Abbreviations are introduced in parentheses where they first appear in the text.

#### 1. Echocardiographic diagnosis

Echocardiography is a usuful non-invasive diagnostic method for cardiovascular lesions, specifically coronary artery lesions due to Kawasaki disease (mucoutaneous lymph node syndrome). Echocardiographic examinations are strongly recommended for patients with Kawasaki disease or with a past history of Kawasaki disease. The following remarks are confined to cursory screening by echocardiography without going into specific detailed examinations.

#### i. Demonstration of the cardiac structures

Two-dimensional echocardiography (2DE) with a sector or linear scanner is required. In order to obtain satisfactory images, use of a high-frequency transducer is desirable. Careful observations of real-time images either on a TV monitor, video recorder or cinematograph are essential.

#### ii. Approach to coronary artery

An example of the standard approach to demonstrate the coronary artery by 2DE is described below as a guideline. As far as the left coronary artery (LCA) is concerned, the left main coronary artery (LM) may be demonstrated on the horizontal plane from the third left intercostal space by obtaining the cross-sectional view of the aortic root. The anterior descending branch (LAD) may be visualized by moving the transducer laterally and rotating the transducer plane slightly counterclockwise. The circumflex branch (LCX) also can be demonstrated basically in the same way. For demonstration of the right coronary artery (RCA), the tricuspid anulus should be visualized first on the horizontal plane from the fourth left intercostal space, and then the probing plane is turned upward (Segment 1). Subsequently, the tricuspid anulus is demonstrated on the sagittal section from the third left intercostal space, and the plane is turned laterally (Segment 2).

#### iii. Coronary artery lesions on 2DE

All echocardiographic records should have notations of date of examination. Coronary artery lesions (CAL) are divided into dilated lesions (DL) and stenotic lesions (SL). For description of DL, definite lesions should be described as df DL, but df can be omitted for the sake of simplicity. For lesions which are not obvious, but strongly suspected, description

"suspicious of" (s/o) DL can be used. If DL cannot be ruled out, but is unlikely, it can be described as "rule out" (r/o) DL. If one must categorize patients according to eithter presence or absence of DL, s/o DL should be included in the group with DL, and r/o DL should be disregarded.

Although quantitative measurement of DL is desirable, sufficient data on the echocardiographic measurements of the normal coronary arterial size are not yet available at this time. For the time being, the following arbitrary criteria should be employed. Diagnosis of DL should be made first by carefully comparing the internal diameter of the coronary artery to that of the adjacent portion of the vessel, and by considering the age of the patient; as well as the possibility of a difference of diameter between right and left coronary arteries (so-called dominance). Changes in the coronary artery diameter during the course of disease should be taken into account. The internal diameter of coronary artery should be measured on a 2DE image of good quality and for the sake of uniformity is defined as the inside-to-inside distance between opposing wall echo-s. An increase of coronary internal diameter to more than 1.5 times the adjacent vessel diameter should be considered DL. For children under 5 years of age, coronary arteries with diameter of 3 mm or greater should be regarded as DL. DL is subclassified into large aneurysms (AN1), medium-sized aneurysms (ANm) and small aneurysms (ANs) (or dilatation—Dil). Angiographic criteria described below should be applied to the echocardiographic diagnosis of those subcategories. The location of DL should be described in the same way as the angiographic examination (American Heart Association segment classification). Recording the size and location of DL in a rough sketch such as shown in Fig. 1 is recommended.

The severity of DL may change in the course of disease. It is generally accepted that DL first appear on 2DE around the 9th day of illness on the average, and may diminish in size or sometimes disappear completely on 2DE in the course of illness.

In diagnosing DL by 2DE, false-positive findings are relatively rare. However, 2DE can miss some DL. Such cases with false-negative examination correspond to only minor DL (ANs or Dil) in the peripheral portion of the coronary artery. As far as false-positive findings are concerned, structures which can be mistaken for DL include the atrial appendages, particularly the left atrial appendage. Good understanding of coronary artery anatomy and their relationships with surrounding structures is essential. Continuity of lumen from the aortic root to the coronary artery via the coronary ostium should be confirmed.

It is often difficult to diagnose SL on the basis of 2DE alone. However, many patients with SL have associated DL as well. Therefore, the majority of patients with SL can be detected by 2DE screening. But in a few exceptional patients with history of Kawasaki disease several years prior to the 2DE examination, SL may be present while DL may no longer be detectable. In such patients, 2DE may not be sufficient enough for assessment of the cardiovascular system. If one needs to categorize patients with DL in terms of a single representative lesion, DLs should be ranked in the following descending order of importance: ANI, ANm, and ANs (or Dil).

In addition to DL and SL, increased echo density (ID) of the coronary artery walls or surrounding areas and abnormal thrombus-like mass echos (ME) should be recorded if present.

#### iv Other abnormalities on 2DE

Pericardial effusion (PE) may be observed in the acute phase. The amount of PE is divided into 3 grades: large (l), moderate (m) and small (s).

As far as the ventricular wall motion is concerned, decreased contractility (DC) is defined when the left ventricular ejection fraction(LVEF) is decreased below 60%. Attention must be paid to any ventricular aneurysm (VA) as well as to regional dyskinesis (DK). Mitral, aortic or tricuspid regurgitation (MR, AR or TR, respectively) may occur transiently or permanently. The valvular abnormalities may be evident on 2DE, but pulsed Doppler interrogation is very helpful for assessment of valvular lesions. Sometimes, mitral valve prolapse (MVP) is seen in association.

In patients with CAL, careful search for an aortic aneurysm (AOAN) and for peripheral arterial aneurysms (PAAN), such as aneurysms of the subclavian and iliac arteries, is recommended. Gallbladder distension (GBD) is occasionally seen by 2DE.

When no cardiovascular abnormalities are noted on 2DE, such patients should be described as normal (N). If there is any abnormality besides the lesions described above, it should be described precisely.

#### 2. Angiocardiographic diagnosis

Angiocardiography (ACG) is recommended in patients with clinical evidence of cardiovascular complications including abnormalities on 2DE. The following remarks are concerned only with the regular routine examinations.

### i. Angiographic technique

For coronary angiography (CAG), cineangiography, especially selective coronary angiography (SCAG), is preferred. Multiple views are necessary and occasionally cranial tilt is useful.

# ii. The diagnosis of CAL by CAG

In recording CAG findings, the interval from the onset of the disease should be specified. The names of coronary arteries and branches to be used in a CAG report should be the same as in 2DE; i.e. LCA, LM, LAD, LCX and RCA. CAL is also classified by CAG into two major categories: DL and SL. Quantitative evaluation of DL is recommended, although a complete data-base of normal values in children is not available. Therefore, for the time being, the diagnosis should be based on the following arbitrary criteria. A large aneurysm (ANI) is defined as dilatation with an internal diameter of more than 4 times that of the adjacent vessel diameter; a moderate aneurysm (ANm), between 1.5 and 4 times; and a small aneurysm (ANs), less than 1.5 times. One may call ANs "dilatation" (Dil). Small dilatation of the bifurcation of LCA may be called a "web" (Web).

The location of DL should be expressed by the segment (Seg) of the American Heart Association (AHA) classification (Fig. 2). For example, DL located in Seg 1 is expressed "Seg 1"; DL involving Seg 1 and Seg 2 is expressed "Seg 1-2", and DL involving the junction between Seg 1 and Seg 2 "Seg 1×2". DL at the bifurcation of Seg's 5, 6 and 11 (bifurcation of LM into LAD and LCX, which is the most frequently involved area) is expressed "Seg 5×6". DL frequently develop at branching regions of coronary arteries.

SCAG is highly recommended for assessment of SL. SL is subclassified into 3 groups: occlusion (OC), segmental stenosis (SS) and localized stenosis (LS). OC is defined as complete obstruction of the coronary artery, and SS is defined as severe narrowing over a long segment of the coronary artery. SS is considered to be a result of recanalization of a throm-

bosed aneurysm. LS may be further described in terms of percent narrowing of the lumen diameter such as: 25, 50, 75, 90 and 99%, or in terms of grade or severity, such as high (hg), moderate (md) and slight (sl). Roughly, 99% corresponds to hg; 90–75% to md, and 50–25% to sl. When the degree of stenosis cannot be specified, it should be expressed "undetermined" (ud).

The location of SL also should be expressed according to the AHA segment classification (Fig. 2). SL very rarely develop in branching portions of the coronary artery. OC and LS should be described specifying the involved segment. SS should be described enumerating all the segments involved, e.g. Seg 1–2–3, indicating that SS ranges from Seg 1 through Seg 3.

If one needs to categorize patients according to a single most important lesion, the following order of priority should be used: OC, SS, LS and AN, although this order does not necessarily correlate with the clinical significance.

Collateral circulation (CL) of the coronary artery, if observed, should be recorded. CL is subclassified into 2 groups: intracoronary collateral circulation (intra-CL) and intercoronary collateral circulation (inter-CL). Whenever plausible, the term SS should be used rather than intra-CL.

Irregularity (IR) of the coronary arterial wall, an intracoronary thrombus-like filling defect (TH), and abnormal tortuosity (TT) of the coronary artery should also be recorded, but TT should not be used in the description of SS or CL. In order to illustrate the nature and the extent of CAL, the diagram shown in Fig. 2 is recommended.

Beside CAL, if there is any discrepancy in the size of coronary arteries between right and left, the dominant vessel should be specified. In addition, any congenital abnormalities of the coronary artery should be described.

# iii. Diagnosis of cardiovascular lesions other than CAL

Aortic aneurysm (AOAN), periperal arterial obstruction (PAO), stenosis (PAS), and aneurysm (PAAN) should be recorded if present. When left ventricular ejection fraction (LVEF) is decreased to less than 50%, it should be noted as "decreased contractility" (DC). The other findings such as regional dyskinesis (DK) or ventricular aneurysm (VA) should be described.

As far as cardiac valvular abnormalities are concerned, findings which should be noted include aortic regurgitation (AR), mitral regurgitation (MR) and mitral valve prolapse (MVP).

Pulmonary hypertension (PH), if present, should be recorded along with the pressure value. Also, pulmonary arterial wedge pressure and left ventricular end-diastolic pressure should be recorded as appropriate.

Calcification (CC) of the coronary artery or epicardium is occasionally observed. It may not appear clearly on regular chest X-rays, but only on fluoroscopy or cineangiography. Any calcication of the cardiac structures should be recorded. When CC is present in the extracoronary structures, it should be described as extra-coronary calcification (extra-CC). When CC of the coronary artery is noted, the location of CC should be described in such a way as RCA-CC and LAD-CC.

When no abnormalities are found in either ACG or cardiac catheterization studies, the patient is regarded as "normal" (N). If there is any abnormality (on any investigation,) besides the findings described above, it should be precisely recorded.

#### **APPENDIX**

# LIST OF ABBREVIATIONS

2DE Two-dimensional echocardiography

LCA Left coronary artery

LM Left main coronary artery

LAD Left anterior descending branch

LCX Left circumflex branch
RCA Right coronary artery
CAL Coronary artery lesion(s)

DL Dilated lesion(s)
SL Stenotic lesion(s)

df Definite

s/o Suspicious of

r/o Rule out
AN Aneurysm

ANI Large aneurysm

ANm Medium-sized aneurysm

ANs Small aneurysm

Dil Dilatation

ID Increased echo density (of coronary arterial walls)

ME Thrombus-like mass echos

PE Pericardial effusion

PEI Large pericardial effusion
PEm Medium pericardial effusion
PEs Small pericardial effusion

DC Decreased contractility (of the left ventricle)

LVEF Left ventricular ejetion fraction

VA Ventricular aneurysm
DK Regional dyskinesis
MR Mitral regurgitation
AR Aortic regurgitation
TR Tricuspid regurgitation
MVP Mitral valve prolapse
AOAN Aortic aneurysm

PAAN Peripheral artery aneurysm

GBD Gallbladder distension

N Normal

ACG Angiocardiography
CAG Coronary angiography

SCAG Selective coronary angiography
AHA American Heart Association

OC Occlusion

SS Segmental stenosis
LS Localized stenosis

hg High (degree or grade)

md Medium sl Slight

ud Undetermined

CL Collateral circulation

intra-CL Intracoronary collateral circulation inter-CL Intercoronary collateral circulation

IR Irregularity of the coronary arterial wall

TH Thrombus-like filling defect

TT Abnormal tortuosity of the coronary artery

PAO Peripheral artery obstruction
PAS Peripheral artery stenosis

CC Calcification

extra-CC Extra-coronary calcification

Fig. 1:An example of illustrating echocardiogram of the coronary artery

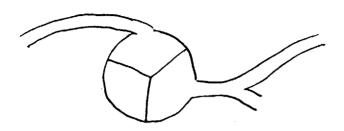


Fig. 2: Classification of the segment of the coronary artery (AHA)

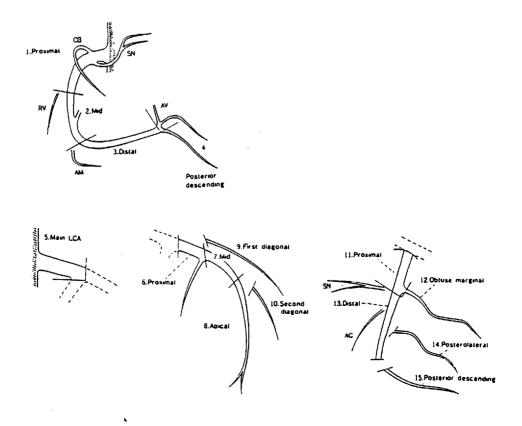


Fig. 3: An example of normale echocardiogram of the left coronary artery

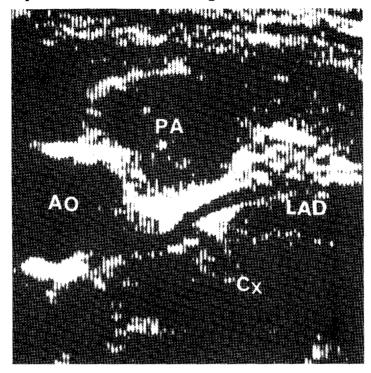


Fig. 4: An example of normale echocardiogram of the right coronary artery

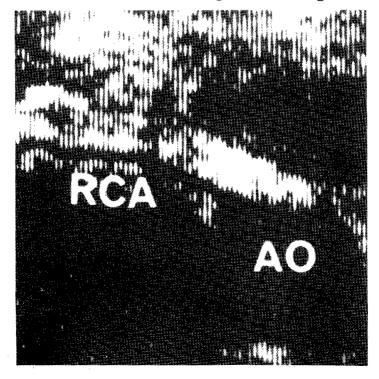


Fig. 5: Web. Seg  $5 \times 6$ .

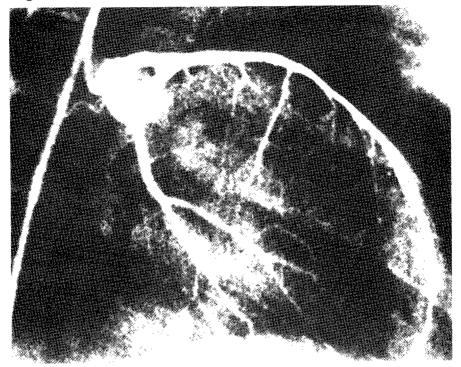


Fig. 6: LSmd, Seg 6. ANm. Seg.6-7.

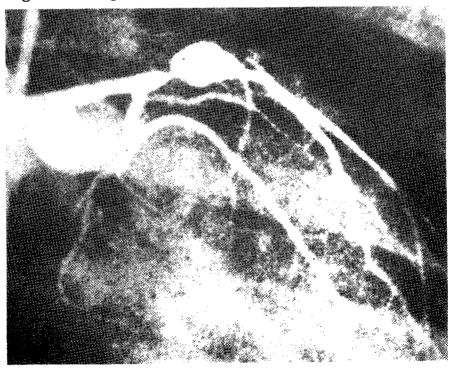
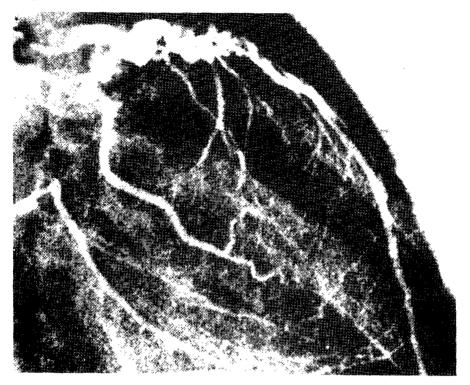


Fig. 7:0C, Seg 13. CL. intra. SS, Seg 6-7. ANs. Seg 5-6. ANs, Seg 11-12.



# 検索用テキスト OCR(光学的文字認識)ソフト使用 論文の一部ですが、認識率の関係で誤字が含まれる場合があります

本委員会は、昨年度に"川崎病による心血管障害の診断基準"をすでに報告したが、本年度は、この基準の英訳をおこなった。英文は Los Angeles の Dr, Takahashi、Chicago の Dr. Schulman の手をわずらわせた。記して感謝する。