Coronary Artery Involvement in Children With Kawasaki Disease: A Pediatrician’s Dilemma.

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Introduction

Kawasaki disease (KD) is an acute, self-limiting vasculitis of unknown etiology that affects predominantly infants and young children. The entity was first described by Tomisaku Kawasaki in 1967 and it was Dr Takajiro Yamamoto who first noted its cardiac involvement in non-fatal cases. The diagnosis of KD is made clinically. There is currently no diagnostic test for this disease. Clinical criteria aid clinicians in making the diagnosis of KD.

The presence of coronary artery aneurysm is the most severe outcome in KD, but even the transitory dilatation of the coronary arteries is important in the diagnosis, management, and follow-up of children with KD. The identification of coronary artery dilatation at onset of KD has become pivotal in supporting the diagnosis and guiding further management.

Case Report

A 4 year old boy, completely immunized and developmentally normal for age, with a weight of 12 kg and 96 cm tall presented with 5 days history of high grade fever and maculopapular rashes over the trunk, anterior aspect of thigh and arms. Red lips with no cracking was noted, but no lymphadenopathy, edema, peeling or conjunctival injection. Histories of atopy or drug intake were negative. All differentials of viral exanthematous fevers were excluded and possibility of staphylococcal infection/ Kawasaki disease was considered. All investigations were normal except for the total count of 15000/mm³ and ESR of 48 mm/hr. Platelet count was normal throughout the course of illness. Echo done on day 7 of illness showed LMCA (Left main coronary artery) 2.8 mm and RCA (right coronary artery) 2.3 mm and without other abnormalities. Child was started on anti-staphylococcal antibiotics but fever persisted, and there was no clinical improvement. A repeat echo was done on day 11 showed LMCA and RCA having the same dimensions.

A normogram referring to the coronary artery dimensions in children was referred to and the Z scores for this child as per the body surface area (BSA=0.56 m²) is as follows (in millimeters)
LMCA: Z score: (0Z= 1.99 , 1Z=2.35 2 Z= 2.72 and 2.5 Z=2.91)
Proximal RCA= Z Score: (0Z = 1.74, 1Z= 2.11 2Z= 2.49 and 2.5Z = 2.68)
LAD Z score: (0Z= 1.53, 1Z= 1.87 2Z= 2.21 and 2.5Z= 2.38)
Accordingly LMCA is > 2 Z scores as per normogram and could be inferred as dilated.
Child was given IVIG and Aspirin and recovered gradually. A repeat echo at discharge showed the dimensions are reduced to less than 1 Z score
Here our dilemma was
(1) How to interpret the measurements of coronary artery dimensions as per different age groups in echocardiogram to state whether they are dilated or not?
(2) How reliable are LMCA measurements compared to RCA and LAD in further management?

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References

Abbreviations used in the article

IVIG = Intravenous Immunoglobulin
Erythrocyte Sedimentation Rate
RCA = Right Coronary Artery
LCX = Left Circumflex Coronary Artery
LAD = Left Anterior Descending Coronary Artery
LMCA = Left Main Coronary Artery

Discussion

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Commonly used criteria of coronary artery (CA) involvement have relied on the Japanese Ministry of Health criteria, which dichotomously define abnormalities as a maximum absolute internal diameter > 3 mm in children less than 5 years of age or >4 mm in children, 5 years and older, or a segment 1.5 times greater than an adjacent segment, or the presence of luminal irregularity. This criterion contains subjective variations, does not account for variations according to body surface area, and ignores any continuum or time course.

Normogram for coronary artery dimensions have proven to be an essential aid in predicting the course, and in the decision of precise therapeutic intervention. The intraluminal diameters of CA segments are measured from inner to inner edge. CA and aortic valve diameters are expressed in millimeters, weight in kilograms, height in centimeters, and BSA in square meters. In order of descending frequency, common sites of coronary aneurysms include the proximal LAD and proximal RCA, followed by the LMCA, then LCX, and finally the distal RCA and the junction between the RCA and posterior descending coronary artery.

For the purposes of analysis, measures of the z scores of the proximal LAD and proximal RCA branches are used. Normal values do not currently exist for distal segments or the circumflex branch. Although normogram exists for the LMCA, its measurements, with relation to anatomic variations make its interpretation less reliable, and it is exceedingly rare to have enlargement of the left main coronary artery in KD without accompanying dilation of the proximal LAD. The definition of coronary artery involvement has important implications for follow-up, and in the identification of risk factors which may distinguish the high-risk patients who might require serial evaluation and more aggressive treatment.

Conclusions

If Z scores become the basis of the CA dilatation definition, they must be derived from rigorous models with appropriate validation to avoid misclassification. BSA- adjusted coronary artery dimensions on baseline echocardiography is essential in health care in general, allowing appropriate predictions of disease and thus better therapeutic intervention.

References


Abbreviations used in the article

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<tr>
<th>Abbreviation</th>
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<td>BSA</td>
<td>Body Surface Area</td>
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<td>CA</td>
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<td>KD</td>
<td>Kawasaki Disease</td>
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<td>LMCA</td>
<td>Left Main Coronary Artery</td>
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<td>LAD</td>
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<td>ESR</td>
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