# [Examining the Utility of Coronary Artery Lack of Tapering and Perivascular Brightness in Incomplete Kawasaki Disease.](https://www.ncbi.nlm.nih.gov/pubmed/30196380)

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**Take Home Points:**

* **Lack of tapering of the coronary arteries (LT) and perivascular brightness (PB) are subjective, poorly reproducible, and not specific for Kawasaki disease (KD)..**
* **Guidelines should ideally be based on data and consensus panel recommendations should be subsequently tested for validity when possible.**



**Commentary from Dr. Jared Hershenson (Greater Washington DC), section editor of Pediatric Cardiology Journal Watch: In 2017, the AHA published revised guidelines for the diagnosis of KD, including more extensive recommendations for incomplete KD. Both LT, defined as absence of the normal tapering expected in the distal coronary, and PB, defined as echogenicity/brightness of the coronary wall, which had been used in the previous guidelines, were removed due to the absence of data, with the only “suggestive of KD” coronary artery criteria being 2D measurements of at least a z-score of 2-2.5. The inclusion of these criteria were initially based on expert panels. The thought was that LT or PB may represent arteritis prior to true dilation or aneurysm formation.**

**This was a single center retrospective study that began prior to the new revisions. They hypothesized that both LT and PB were unreliable. 117 de-identified parasternal short axis echo clips of the coronary arteries (CA) from patients ages 0-10 years old were interpreted blindly by 6 pediatric cardiologists with at least 5 years of experience outside of training. Subjects were grouped as 1) healthy, afebrile, with benign murmurs, 2) KD s/p IVIG treatment, 4-5 clinical criteria at presentation based on chart review, 3) incomplete KD (iKD) s/p IVIG, 1-3 clinical criteria, 4) febrile > 3 days, no IVIG, KD not suspected. The KD and iKD patients were identified though a list of all patients receiving IVIG and subsequent chart review for symptoms and diagnoses. Each of the 6 reading cardiologists interpreted the same clips independently and recorded the presence or absence of LT and PB, and measured the diameters of the LMCA, LAD and RCA. At least 10 duplicate cases were selected to assess intra-observer reliability. Since the reading cardiologists were blinded, they could not calculate z-scores based on the measurements obtained.**

Median ages of the groups ranged from 1.7-3.2 years. Chart review showed subungual peeling and/or CA aneurysm (z-score > 2.5) were noted in 21/30 KD and 19/32 iKD patients. CA z-score measurements showed good intra-observer agreement (reliability coefficients 0.52-0.6). In contrast, LT and PB showed poor agreement (reliability coefficients 0.36 and 0.13 respectively). See table 2. The prevalence of positive LT and PB in the healthy and febrile groups outnumbered the total in the KD and iKD groups from some of the echo readers, with the median frequency for LT 11% healthy, 53% KD, 44% iKD, and 24% febrile groups, and for PB 4% healthy, 17% KD, 14% iKD, 11% febrile groups. The rate of agreement for positive LT reads in a majority of cardiologists (e.g. 4-6) was very low. For PB, in 92% of the patients there was good agreement for a negative read, and only a single patient had a majority agreeing on a positive read (see figure 1). The intra-observer variability was low for both LT and PB (see Table 2). At least one cardiologist had a high percentage of LT and PB on all clips, suggesting “over-diagnosis” likely due to the unreliability of these findings. Review of the medical records showed that the presence of LT and/or PB played a role in the diagnosis of iKD and subsequent IVIG treatment in some patients. One of the four did have periungual peeling and none had late aneurysms. Additionally, none of the 10 iKD patients with LT or PB had refractory symptoms requiring another dose of IVIG or other medication.

Limitations of this study included its retrospective nature and a statistical difference in age between the groups. However, this study showed that LT and PB are poorly reliable, poorly reproducible, and not specific to iKD or KD. Additionally, these findings were seen in normal and febrile patients without KD, a known issue amongst cardiologists who read many echocardiograms for suspected KD (usually iKD) without enough clinical criteria to meet even incomplete diagnostic criteria, but with the pediatricians or ID folks hoping we can help them “make the diagnosis”. As the authors note, guidelines without evidence can often be flawed. There should be future prospective studies especially in KD looking at the utility of the current guidelines and use of echocardiography in the diagnosis.



