

Visual assessment of restricted left atrial motion: significance in differential diagnosis and prognosis in patients with cardiac amyloidosis and hypertrophic cardiomyopathy

Haruhiko Higashi, Katsuji Inoue, Chiharuko Iio, Tamami Kono, Teruyoshi Uetani, Jun Aono, Takayuki Nagai, Kazuhisa Nishimura, Jun Suzuki, Takafumi Okura, Jitsuo Higaki, Shuntaro Ikeda

Department of Cardiology, Pulmonology, Hypertension & Nephrology, Ehime University Graduate School of Medicine, Toon, Japan

Background: Because the two-dimensional echocardiographic findings of cardiac amyloidosis (CA) mimic those of hypertrophic cardiomyopathy (HCM), the differentiation of these two entities can be still challenging.

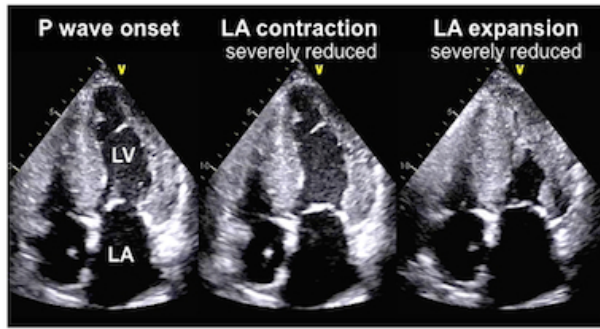
Purpose: This study aimed to evaluate the significance of visually assessed left atrial (LA) motion in differentiating CA from HCM and predicting prognosis in these patients.

Methods: A total of 124 HCM patients (mean 64 years, 29% in female) and 33 biopsy-proven CA patients (mean 71 years, 12% in female) were retrospectively studied. Echocardiography was performed during sinus rhythm at the first visits to our institution from 2006 to 2015. Visual judgments of global LA motion were made from apical four-chamber view. The inward motion during atrial systole (LA contraction) and the outward expansion during ventricular systole (LA expansion) were semiquantitatively scored as follows: (1) normal, (2) moderately reduced, and (3) severely reduced or akinetic. LA wall motion score index was calculated as the averaged of the scores. In addition, two strain parameters (LA booster strain and total reservoir strain) were measured using speckle tracking. Late gadolinium enhancement with cardiac magnetic resonance imaging (CMR) could be evaluated in 97 HCM patients and 21 CA patients.

Results: LA wall motion score index was higher in patients with CA than that in HCM (CA: 2.5 ± 0.6 , HCM: 1.6 ± 0.7 , $P < 0.01$), although there was no significant difference of LA volume index between 2 groups (CA: 50 ± 23 ml/m², HCM: 43 ± 16 ml/m², $P = 0.17$). Figure showed the comparisons between LA wall motion score index, and booster and reservoir strains. The receiver-operating characteristics analysis to differentiate CA from HCM showed that LA wall motion score index ≥ 2.5 had a sensitivity of 72.7% and a specificity of 81.5%. Among patients who underwent CMR, the late gadolinium enhancement in the ventricular wall was frequently observed in 80 HCM patients (82.5%) and 19 patients with CA (90.5%). In contrast, the enhancement in the LA wall was revealed in 1 patient with HCM (1.0%) and 16 patients with CA (76.2%). During follow-up (HCM: mean 4.4 years, CA: mean 2.1 years), 26 HCM and 18 CA patients developed cardiac events (cardiac death, heart failure hospitalization or atrial fibrillation). LA wall motion score index was higher in HCM patients with cardiac events than those without the events (2.3 ± 0.7 vs. 1.4 ± 0.7 , $P < 0.01$), although it did not reach the statistical significance between CA patients with and without cardiac events (2.6 ± 0.5 vs. 2.3 ± 0.6 , $P = 0.08$).

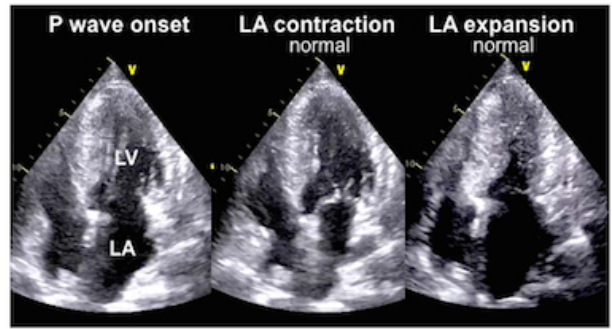
Conclusions: Restricted LA motion occurs frequently in cardiac amyloid patients rather than HCM patients. Risks for cardiac events increased if the restricted LA motion was present in both groups. Vigilant screening for abnormal LA motion might help early recognition of cardiac amyloid patients.

Cardiac amyloidosis (CA)

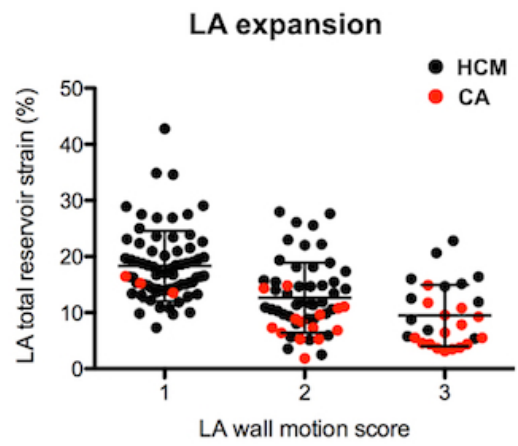
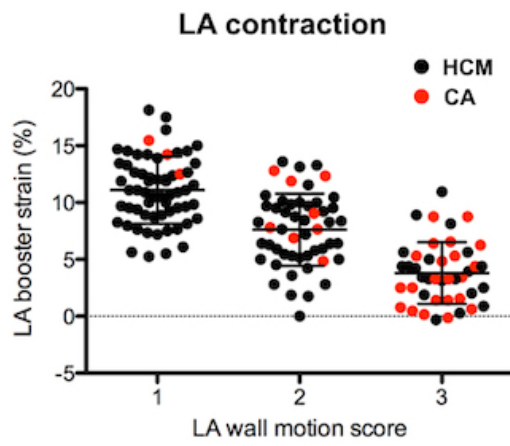


LA wall motion score= 3

Hypertrophic cardiomyopathy (HCM)



LA wall motion score= 1



左房運動の視覚的評価: 心アミロイドーシスと肥大型心筋症の鑑別と予後予測における有用性

東 晴彦、井上勝次、飯尾千春子、河野珠美、上谷晃由、青野 潤、永井啓行、西村和久、鈴木 純、大蔵隆文、檜垣實男、池田俊太郎

愛媛大学大学院医学系研究科 循環器・呼吸器・腎高血圧内科学

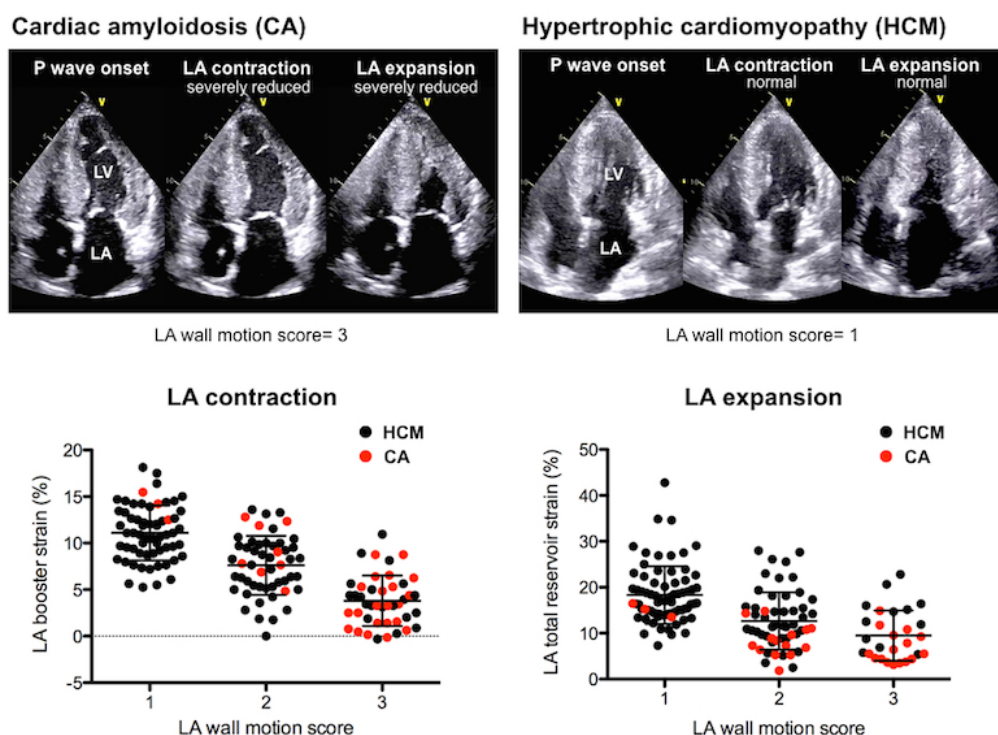
【背景】心アミロイドーシスの心エコー図所見は肥大型心筋症と類似しているため両者を鑑別することは困難なことが多い。

【目的】本研究では、視覚的に評価した左房の動きが心アミロイドーシスと肥大型心筋症の鑑別と予後予測に関して有用か否かを検討した。

【方法】当院を受診した 124 名の肥大型心筋症(平均 64 歳、女性 29%)と 33 名の生検で確定診断された心アミロイドーシス(平均 71 歳、女性 12%)を対象に後方視的に検討した。洞調律時の心尖部四腔断面像で左房の動きを評価した。左房の動きは、内向きの動き(LA contraction)と外向きの動き(LA expansion)をそれぞれ視覚的に 1 点(正常)、2 点(やや低下)、3 点(著しく低下)に半定量評価し、その平均値を LA wall motion score index (LWMSI)と定義した。さらに、スペックルトラッキング法を用いて LA booster strain と total reservoir strain を計測し、LWMSI と比較した。

【結果】左房容量係数は両群間で有意差を認めなかったが、LWMSI は心アミロイドーシスで有意に高値であった(CA: 2.5 ± 0.6 , HCM: 1.6 ± 0.7 , $P < 0.01$)。図に LWMSI と booster strain、reservoir strain の関連を示す。両疾患を鑑別するための ROC 解析を行うと、LWMSI ≥ 2.5 で心アミロイドーシスを感度 72.7%、特異度 81.5%で鑑別可能であった。フォローアップ期間内に 26 名の肥大型心筋症患者、18 名の心アミロイドーシス患者に心血管イベント(心臓死、心不全による入院、心房細動の発症)が発生した。肥大型心筋症において LWMSI はイベントがあった群でイベントがなかった群より高値であり(2.3 ± 0.7 vs. 1.4 ± 0.7 , $P < 0.01$)、心アミロイドーシス群においてもイベントがあった群で高い傾向にあった(2.6 ± 0.5 vs. 2.3 ± 0.6 , $P = 0.08$)。

【結論】心アミロイドーシスは肥大型心筋症に比較して視覚的に評価した左房の動きは制限されていた。両群において高 LWMSI は心血管イベント発症のリスク因子であった。



質疑応答

質問 1:

LWMSI の再現性はどうか？

応答 1:

内相関係数 (ICC) は検者内 0.86, 検者間 0.77 であり、再現性は良好であった。

質問 2:

心アミロイドーシスで LWMSI が高値になる理由は？

応答 2:

心房へのアミロイド沈着が関与している可能性がある。心臓 MRI を施行し得た心アミロイドーシスでは 8 割近くの症例で心房に LGE を認めた。一方、肥大型心筋症では心房の LGE を認めた症例は少なかったため心房の構造的リモデリングは心アミロイドーシスで進行していたと考えられた。