Cardiac Involvement in Rare Forms of Amyloidosis Assessed Using ^{124}I-Evuzamitide PET/CT

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Introduction

- Diagnosis of hereditary amyloidosis and of cardiac involvement by amyloidosis remain challenging.
- ^{124}I-evuzamitide (^{124}I-p5+14, AT-01) is a novel pan-amyloid radiotracer that accurately detects cardiac involvement in light-chain and wild-type transthyretin amyloidosis.
- However, its ability to detect cardiac involvement in rare forms of amyloidosis has not been studied in detail.

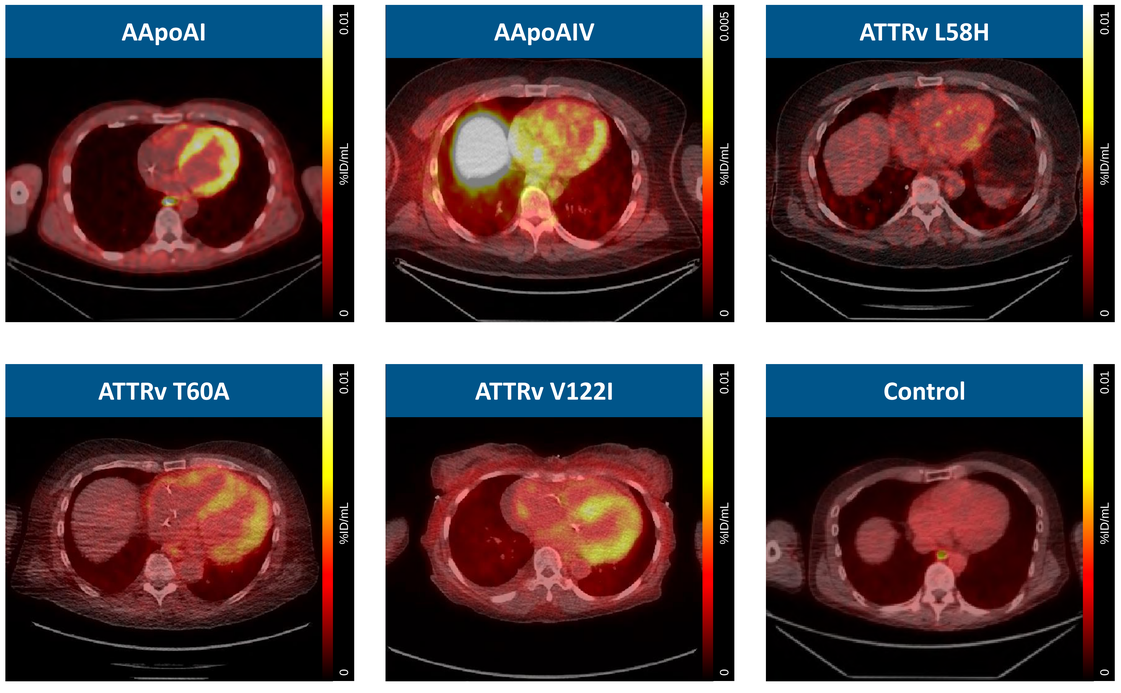
➤ The aim of this study was to compare myocardial uptake of ^{124}I-evuzamitide in rare forms of amyloidosis with or without cardiac involvement and in controls.

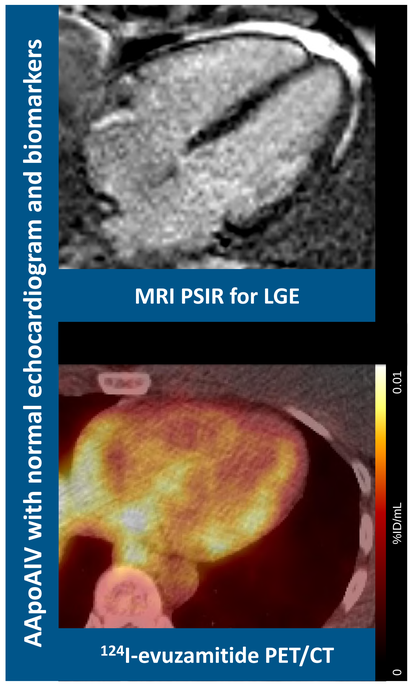
Methods

- We included 20 participants:
  - 7 with hereditary amyloidosis:
    - 2 AApoAI
    - 1 AApoAIV
    - 1 ATTRv L58H (p.L78H)
    - 2 ATTRv T60A (p.T80A)
    - 1 ATTRv V122I (p.V142I)
  - 1 with localized pulmonary light-chain amyloidosis
  - 12 controls without amyloidosis
- Positron emission tomography/computed tomography (PET/CT) was performed 5 hours after administration of ^{124}I-evuzamitide in all participants:
  - Median injected activity 0.97 mCi (IQR 0.93 – 1.10)
- Cardiac scans were assessed visually and quantitatively.
- Myocardial radiotracer activity above the blood pool was quantified as left ventricular percent injected dose:
  - $LV\ \%ID = \frac{mean\ activity\ concentration \times activity\ volume}{injected\ activity}$

Results

- Visual myocardial uptake was present in all hereditary amyloidosis, but absent in localized pulmonary amyloidosis and in all controls.
- Median LV %ID was:
  - 1.6197 (IQR 0.6783 – 2.1057) in hereditary amyloidosis, 0.0001 in localized amyloidosis, and 0.0027 (IQR 0.0007 – 0.0093) in controls ($p < 0.001$).
- For both AApoAI, echocardiogram and cardiac MRI were equivocal for cardiac involvement, but ^{124}I-evuzamitide PET/CT was definitely positive.





Conclusions

- **^{124}I-evuzamitide PET/CT:**
  - **Detects cardiac involvement in multiple forms of hereditary amyloidosis, even when echocardiogram and MRI show no definite involvement.**
  - **Appropriately shows very low myocardial uptake in localized extracardiac amyloidosis and in controls.**

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