Survival in incidentally discovered amyloid cardiomyopathy

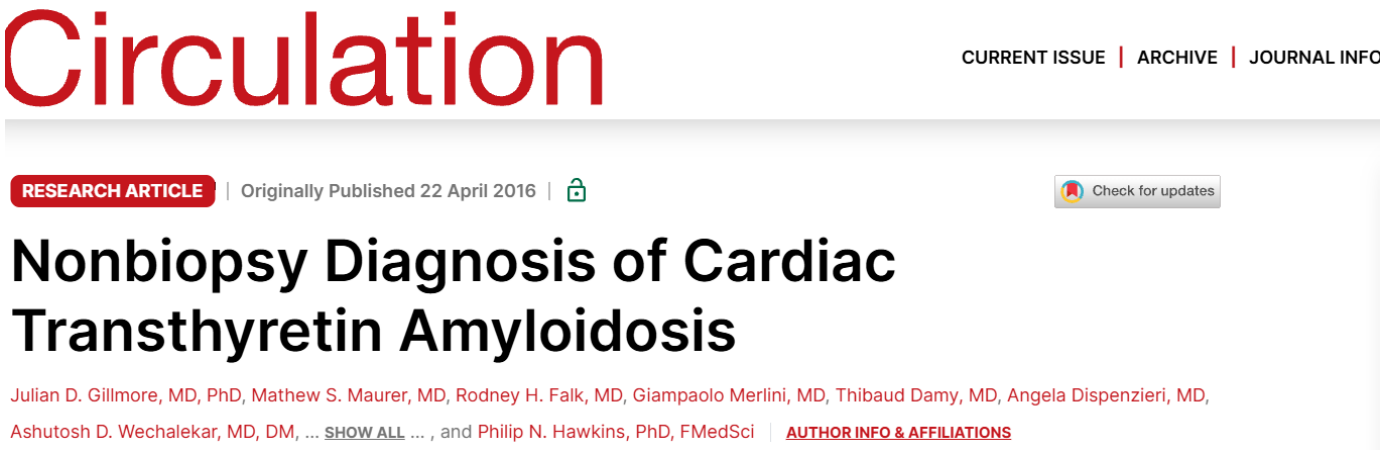
T Bach-Gansmo, S. Ostadi, OMT Thomas, JLG Loaiza, V. Jøraholmen
UNN and AHUS

No disclosures

UNIVERSITETSSYKEHUSET NORD-NORGE
DAVVI-NORGGA UNIVERSITEHTABUOHCCEVIESSU

Cross country skitrack door to door November to May

Transthyretin cardiomyopathy



Circulation

CURRENT ISSUE | ARCHIVE | JOURNAL INFO

RESEARCH ARTICLE | Originally Published 22 April 2016 |

Check for updates

Nonbiopsy Diagnosis of Cardiac Transthyretin Amyloidosis

Julian D. Gillmore, MD, PhD, Mathew S. Maurer, MD, Rodney H. Falk, MD, Giampaolo Merlini, MD, Thibaud Damy, MD, Angela Dispenzieri, MD, Ashutosh D. Wechalekar, MD, DM, ... SHOW ALL ... , and Philip N. Hawkins, PhD, FMedSci | AUTHOR INFO & AFFILIATIONS

- Cardiac uptake of bone tracers have a 98% sensitivity and 86% specificity for transthyretin (ATTR) cardiomyopathy.
- Blood samples ruling out light chain (Kλ) dysglobulinemi is diagnostic (accuracy near 100%) for ATTR
  - With a few, well known differerials

Purpose:

- All patients referred for HDP bone scintigraphy at AHUS during a ten-year period were reviewed to identify the prevalence of cardiac uptake and survival/time to death of these patients
- Analysis included 3168 patients
  - 1801 males median age 75.2
  - 1367 females median age 66.6

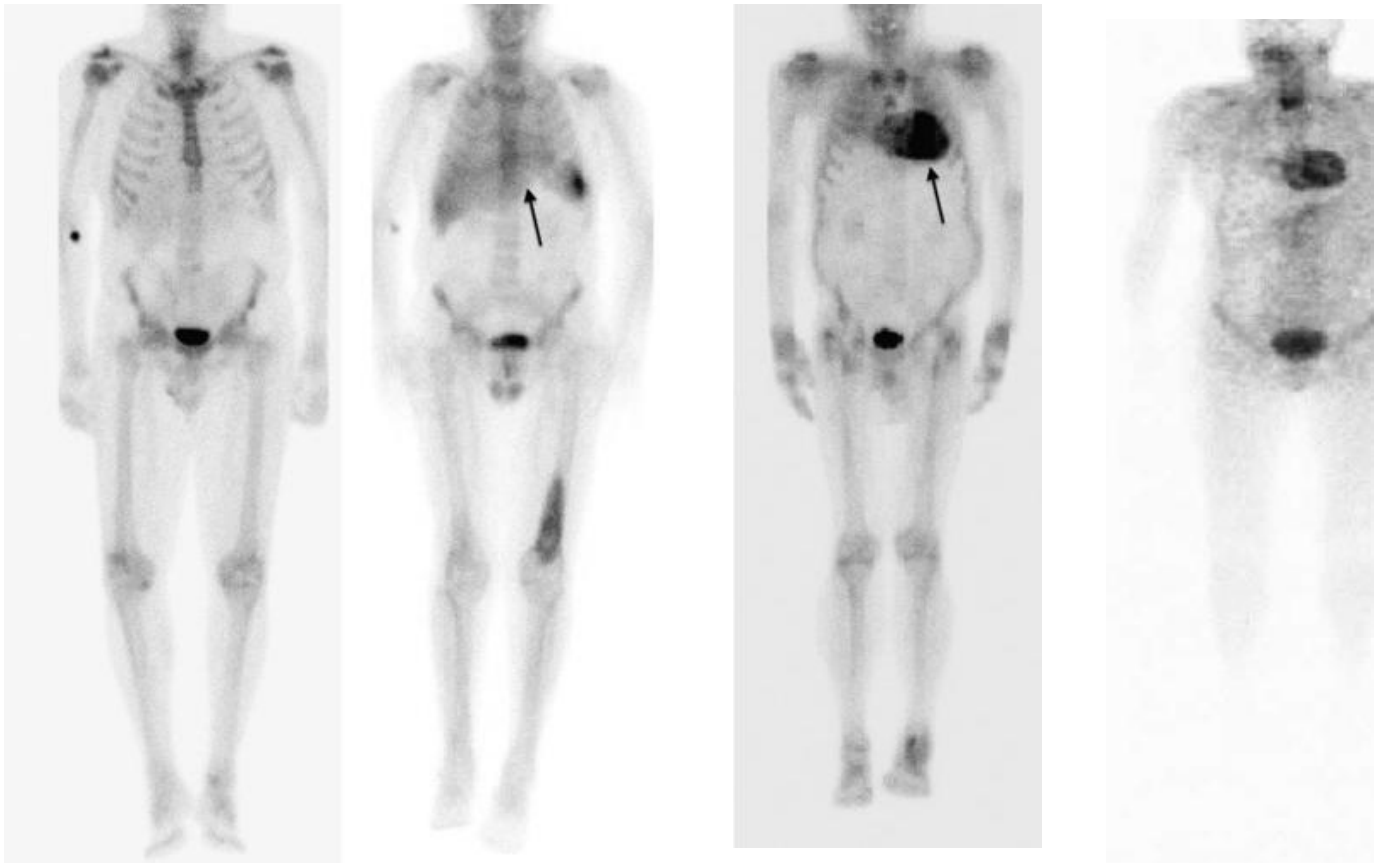
Methods:

All patients above 50 years with HDP bone scan including the thorax were included.

Cardiac uptake was scored according to Perugini.

Perugini score and survival/time to death were recorded.

Censored Kaplan-Meyer data were used to compare Perugini and gender related survival.



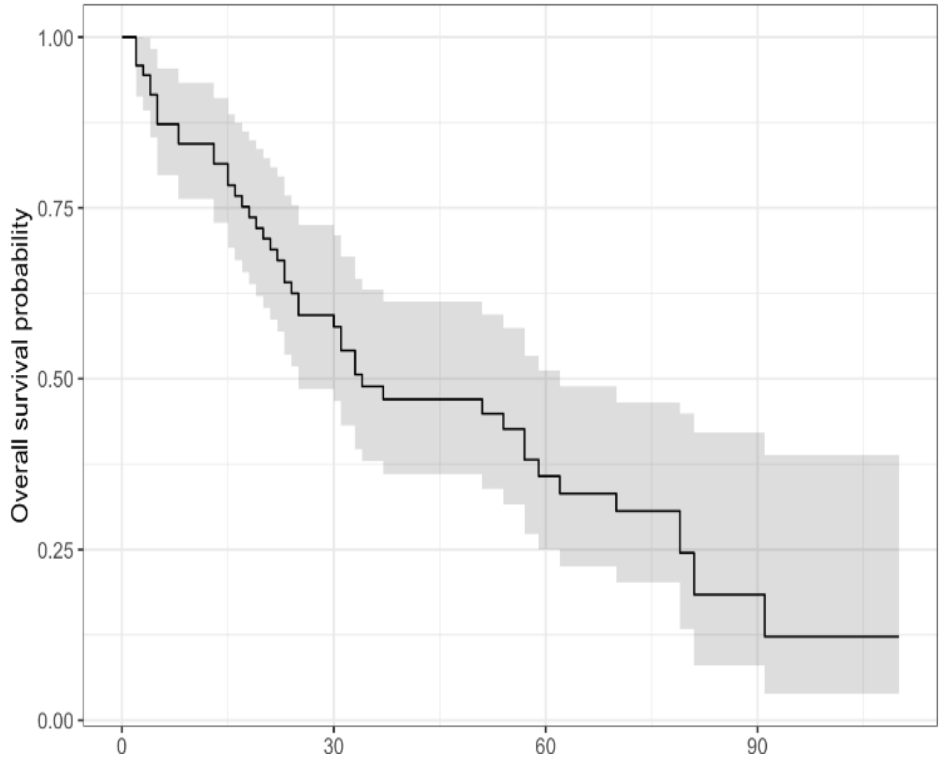
Results:

- 72 patients with cardiac uptake
- 30 patients alive at the end of study.
- Median survival 34 months.
- Censored data show highest mortality in the Perugini 1 group, closely followed by Perugini 3.
- For Perugini grade 1 patients the initial two years after diagnosis was defined by a high mortality, with a subsequent substantial decrease in mortality.

Cardic uptake related to age, gender and mortality.

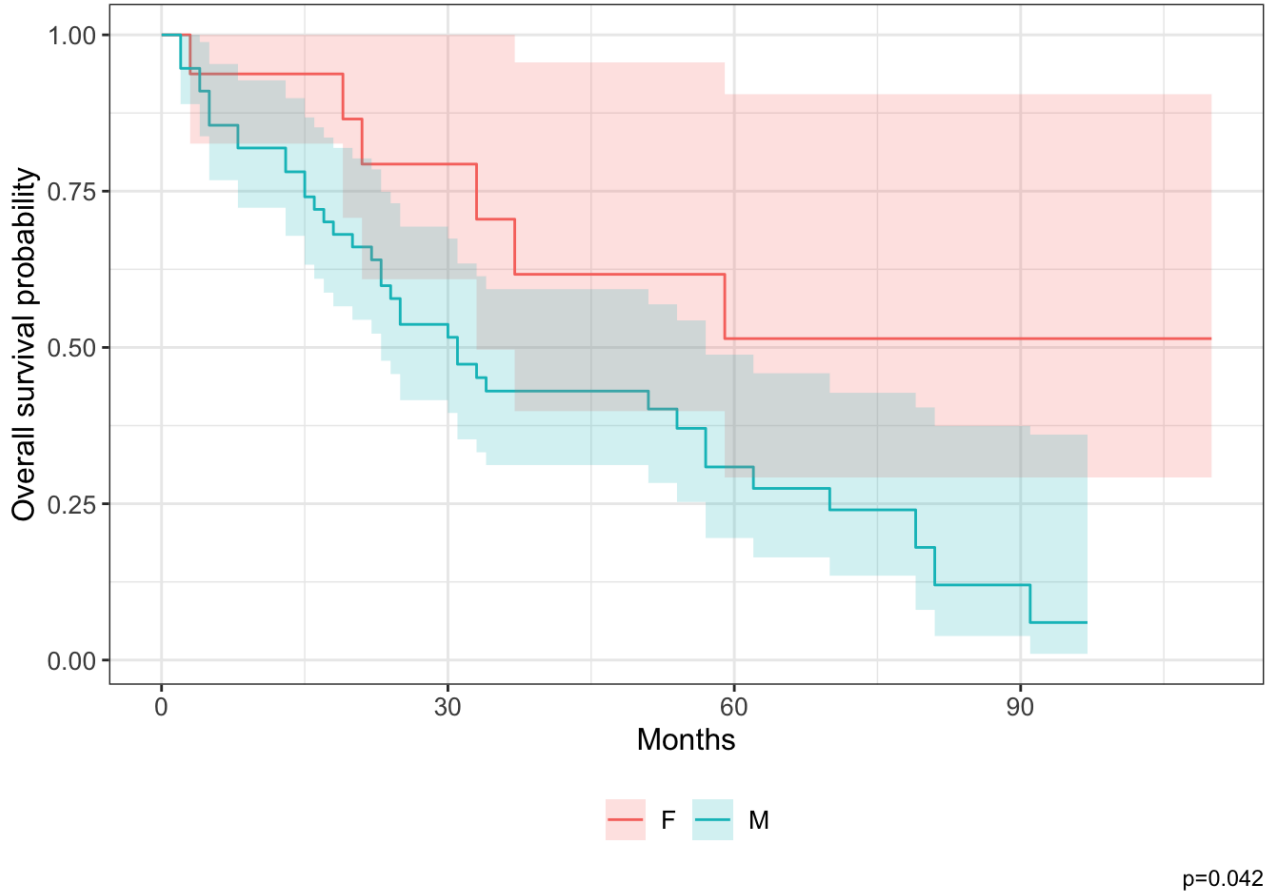
Perugini	1	1	2	2	3	3
Gendre	Male	Female	Male	Female	Male	Female
Dead	20/30	9/14	6/13	1/2	10/14	NA
50-59	0	2	0	0	0	0
60-69	3 1213	2 1145	0	0	1	0
70-79	6	5	1	0	1	0
80-89	19	5	9	2	9	0
90-	2	0	3	0	3	0

Kaplan Meyer curve for all patients
Median survival 34 months



Kaplan Meyer gender related survival.

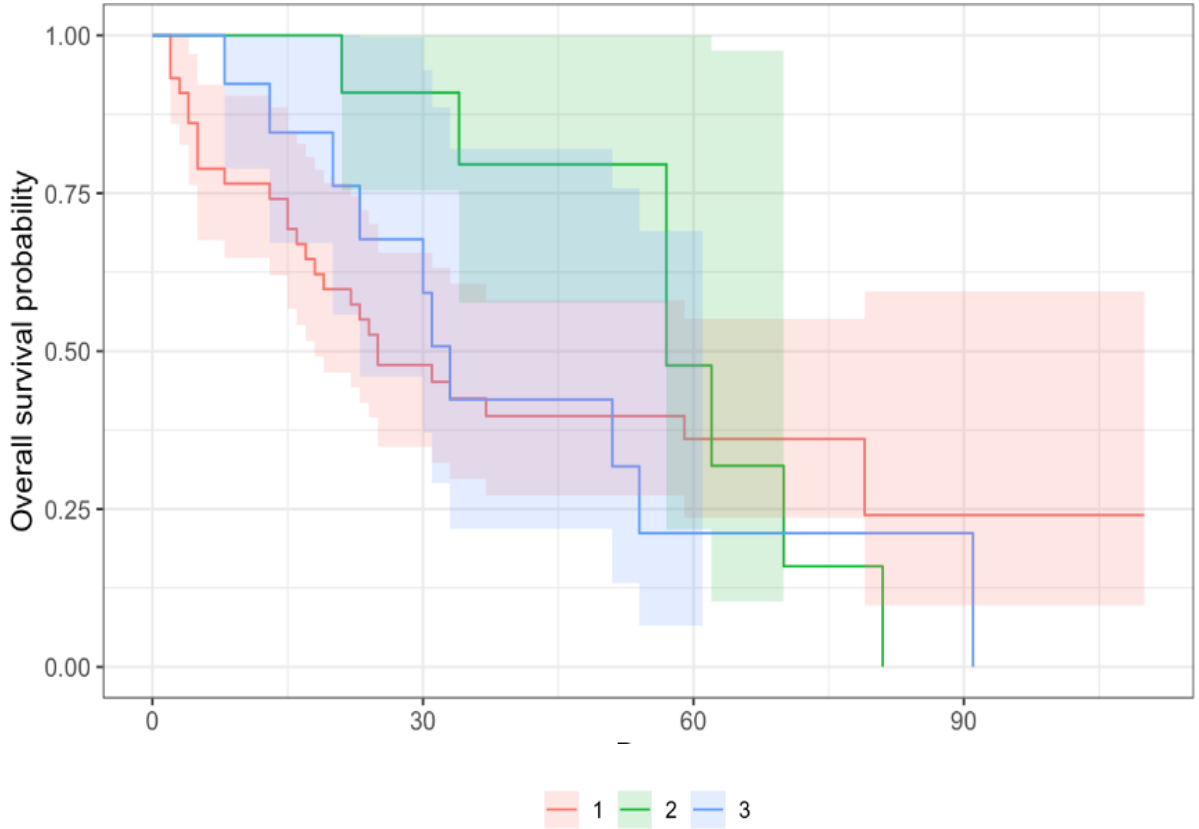
Median survival males 31 months, not reached for females



Median and 25% survival

	50% median survival	75% mortality
All	34 months	79 months males Not reached for females
Perugini 1	28 months	82
Perugini 2	58 months	72
Perugini 3	32 months	52

Kaplan Meyer curve:
Perugini grade related survival



Limitations:

- No SPECT nor biopsy
- Differentlal diagnosis include:

  High bloodpool activity, bone pathology, recent myocardial infarction, hydroxychloroquin toxicity
- No individual had an explanation other than amyloidosis for the cardiac uptake.
  - Eliminate high blood pool uptake or rib fracture as potential cause

Discussion:

- Transthyretin (ATTR) CM is a disease of elderly men, reaching 12% in men above 90 years.
- Perugini grade 3 was not observed in females.
- Amongst patients aged 50-79 years with Perugini grade 1, the incidence was the same for men and women.
- The prognosis was worse for Perugini grade 1 patients than for Perugini grade 2 and 3 patients.

Conclusion:

- Biphasic, high initial mortality
- Highest in Perugini 1 patients
- Similar incidence in males and females below 80 years

- Indicates that, indeed, many of our Perugini grade 1 patients suffered from undiagnosed non-ATTR amyloid deposits, probably immunoglobulin light chain cardiomyopathy.