Extending the Reach of Expert Amyloidosis Care: Development of a Hub-and-Spoke Model

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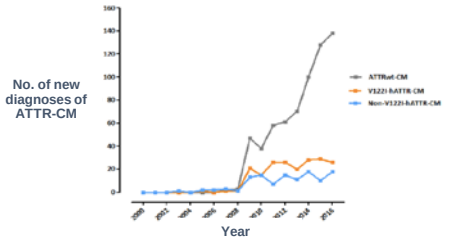
Background: The National Amyloidosis Centre (NAC) was first commissioned in 1999 to provide a diagnostic, staging, monitoring and management advisory service for the national caseload of patients with amyloidosis. Over the last decade, however, there has been a surge in the number of patients diagnosed with cardiac amyloidosis relating to an increased awareness of this disease among physicians, improved diagnostic imaging and the availability of new treatments. In response to this increased demand and in the face of the COVID-19 pandemic, plans are needed for the accelerated development of a UK amyloidosis network.

Increasing number of patients with transthyretin amyloid cardiomyopathy (ATTR-CM)



OBJECTIVES:

The Midlands Amyloidosis Service (MAS) was launched at University Hospitals Birmingham (UHB) in August 2019 with the aim of providing local patients with:

- An early diagnosis
- Virtual, multidisciplinary expertise from the NAC
- Access to novel treatments and / or entry into phase III clinical trials

Emerging Leaders Programme
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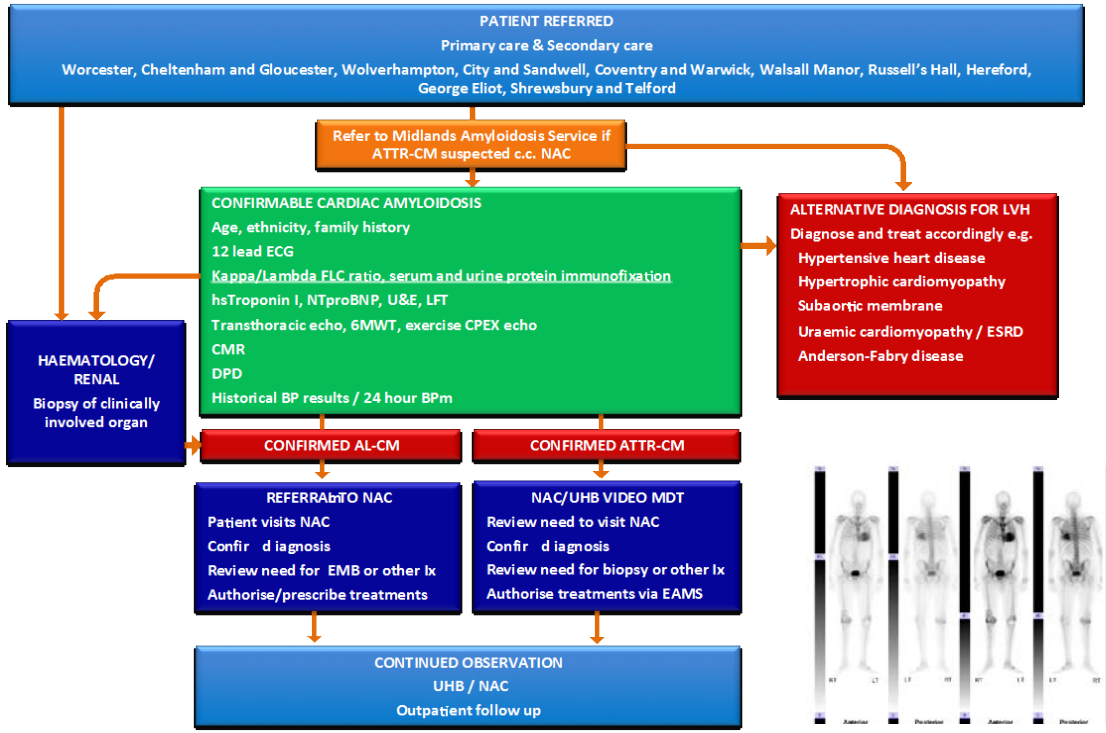
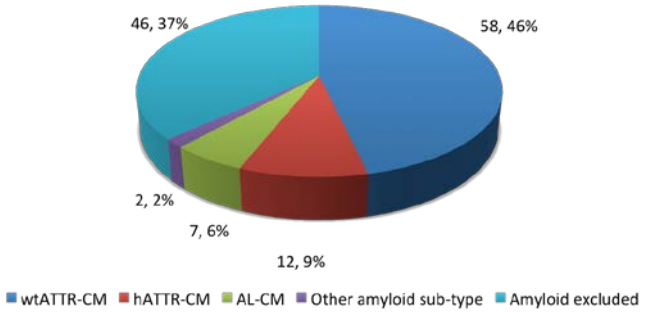
METHODS AND RESULTS:

- A total of 125 patients (age 75 ± 13 yr; male 71%) were referred to the MAS for assessment between 1st August 2019 and 1st January 2021
- All patients were seen in a multidisciplinary clinic with input from consultants in cardiology, nephrology, neurology and genetics
- The median time from referral to diagnosis was 28 days; median time from referral to first clinic assessment was 34 days
- Of the 125 patients referred for assessment 79 (63%) were diagnosed with cardiac amyloidosis; of those, 70 (89%) subjects had transthyretin amyloid cardiomyopathy (ATTR-CM), 7 (9%) had light chain amyloid cardiomyopathy (AL-CM), 1 had (1%) ApoA1 and 1 (1%) had AA amyloidosis sub-types
- Sanger TTR gene sequencing performed at the NAC revealed 12 out of the 70 (17%) patients with ATTR-CM had the hereditary form: V122I (n=7), T60A (n=3) V30M (n=2)
- To date, 50 patients (40%) have been discussed in a video MDT with the NAC, including 14 over the age of 80 years (16%) who had declined to travel to London
- By removing the need for patients to travel to London, a total of 23,207 patient miles were saved (186 ± 28 miles per patient)
- Only two endomyocardial biopsies were required; the majority of diagnoses of ATTR-CM were made possible using the published consensus criteria[1] for the non-invasive diagnosis of wild type TTR amyloidosis
- Of the 58 wild-type ATTR-CM patients, 15 (26%) received tafamidis under the Early Access to Medicines Scheme, and 8 (14%) have thus far been enrolled locally into phase III trials of RNA silencing therapy



Example of 99m Tc-DPD scintigraphy showing grade 2 Perugini uptake - in the absence of a monoclonal dyscrasia these appearances are characteristic for ATTR-CM

Number and Percentage Frequency of Diagnoses After Referral (N =125)



CONCLUSION:

- A hub-and-spoke UK network ensures continued ease and equity of access to specialised amyloidosis healthcare for the increasing numbers of elderly patients diagnosed with ATTR-CM

REFERENCE:

1. Gillmore JD, et al. Circulation 2016;133(24):2404-12