Amyloidosis Workflow

OVERVIEW

Cardiac amyloidosis is a heart condition where protein abnormally builds-up in the heart. This causes heart function to struggle, compensate by working harder -- ultimately resulting in heart failure. There are many causes for cardiac amyloidosis. It can be inherited, triggered by other diseases like cancer, or it may develop on its own. While amyloidosis is not curable, some types are treatable, such as AL (light chain), ATTR, and dialysis-related amyloidosis.

AL amyloidosis occurs when cells in the bone marrow malfunction and produce an excess of light chains. This can happen spontaneously or because of certain types of blood or immune system cancers. Diagnosis of this type of amyloidosis typically happens after age 50.

ATTR Transthyretin (often abbreviated TTR) is a protein that transports a thyroid hormone and vitamin A through the bloodstream. Misfolding of the TTR protein can result in the formation of abnormal protein aggregates called amyloid fibrils in various organs. In the heart, amyloid deposits lead to a condition called senile cardiac amyloidosis.

Corridor4DM provides an Amyloidosis Workflow to allow qualitative, semi-quantitative, and quantitative review of both planar and SPECT patient datasets to assist with identification of cardiac amyloidosis, particularly ATTR.

DATASETS NEEDED

4DM's amyloidosis workflow automatically populates when a Nuclear Medicine (NM) Planar, ungated tomographic SPECT, or a reconstructed tomographic SPECT dataset is loaded, that utilizes either Tc-99m PYP, Tc-99m HDP, Tc-99m DPD, or I-124 Evuzamitide. Both 1-hour and 3-hour protocols can be used and should be noted for inclusion in the report. The radiopharmaceutical should be defined at the camera level. However, when necessary, 4DM’s NM Viewer or MI Processing screens offer an interface—the Dataset Information Window—where you can manually set the dataset's radiopharmaceutical to Tc-99m PYP, Tc-99m HDP, Tc-99m DPD, or I-124 Evuzamitide to activate the amyloid workflow.

HOW-TO-GUIDE

The default amyloidosis workflow layout in 4DM contains five screens for qualitative, semi-quantitative and quantitative review of the dataset. Each screen supports certain NM dataset types. The below sections outline the dataset types required, the purpose of the screen, and the workflow to quantify and grade amyloidosis.

NM Viewer

The NM Viewer screen enables the qualitative, semi-quantitative, and quantitative review of Static (Ungated) Tomographic SPECT datasets. First, for qualitative review, visually compare the tracer uptake intensity between the bone (rib) (See 1, Figure 1) and the heart (See 2, Figure 1) within the dataset display viewports. Assess any uptake in or around the heart.

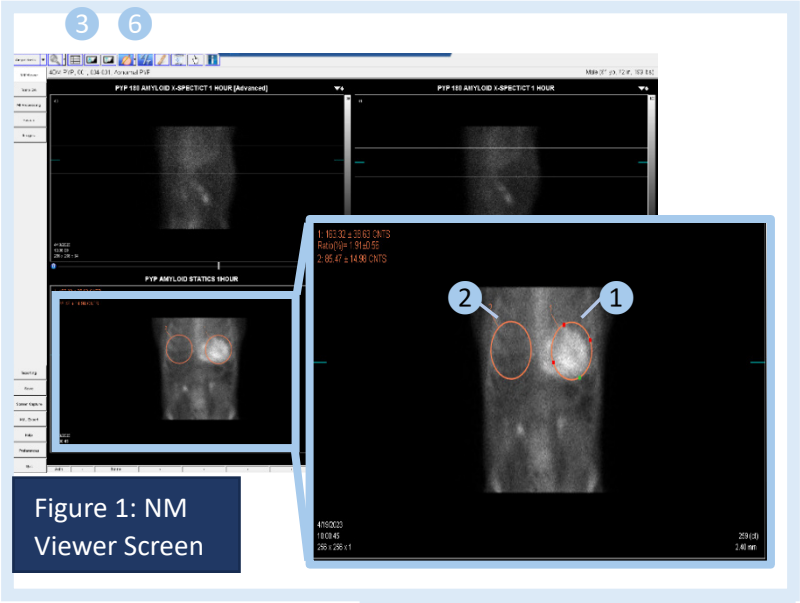

Figure 1: NM Viewer Screen

0Second, estimate a semi-quantitative grade by visual review of the bone and heart uptake, then grading the uptake on a scale of 0-3 in relation to your assessment of heart to rib uptake. See the table below for guidelines.

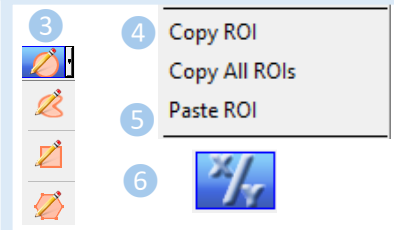

Figure 2: NM Viewer Icons

The final step on this screen is to generate the heart-to-contralateral lung ratio (H/CL). Draw an ROI around the heart using the ROI tool (See 3, Figures 1 and 2). Right click on the ROI and select "Copy ROI" (See 4, Figure 2) then move the cursor to the chest area and “Paste ROI” (See 5, Figure 2). creating an identically sized ROI in the contra-lateral position that is symmetrically positioned to the heart ROI. Selecting the ratio tool (See 6 Figure 1 and 2) generates the H/CL as the ratio of uptake in the heart area to that of the lung area, displaying the H/CL ratio next to the heart ROI statistics.

Guidelines when using Technetium:

Qualitative Assessment	Absent / focal / diffuse
Semi-quantitative grade	0 = no myocardial uptake
	1 = myocardial < rib uptake
	2 = myocardial = rib uptake
	3 = myocardial > rib uptake
H / CL ratio	> 1.6 = suggestive of TTR amyloidosis
	< 1.0 = not suggestive of TTR amyloidosis
	1.0 -- 1.6 = equivocal for TTR amyloidosis

Guidelines adopted from ASNC/AHA/ASE/EANM/HFSA/ISA/SCMR/SNMMI Expert Consensus Recommendations for Multimodality Imaging in Cardiac Amyloidosis

HS_2025-49A

TOMO QA

Visually review the raw planar SPECT data, in the same manner as a TOMO QA review of a myocardial perfusion study (MPI), looking for study artifacts, attenuation patient motion, and overall image quality.

MI Processing

For datasets that are negative, without any uptake in the heart (as evidenced by a semi-quantitative grade of zero, or an H/CL < 1), the MI Processing screen is not necessary to complete the review of the amyloidosis workflow. When the study dataset is positive, perform a typical review of the MI Processing screen, to verify the proper placement of the endocardial and epicardial surfaces. Refer to the MI Processing Help Sheet for guidance on QA steps in this screen.

Fusion

The Fusion screen in the amyloid workflow is only activated when a reconstructed tomographic SPECT dataset acquired with Tc-99m PYP, Tc-99m HDP, or Tc-99m DPD is loaded. This screen enables viewing of both SPECT data and the diagnostic CT or attenuation map from a SPECT or hybrid-CT dataset. Such co-registered datasets are utilized to confirm that the heart uptake is myocardial uptake, not blood pool uptake.

Figure 3: Fusion Screen

To quantify blood pool to myocardial uptake, utilize the following steps:

The VOI display option (See 1, Figure 3) is the default for amyloidosis workflow studies. This option provides volumetric ROI's to compare blood pool to myocardial activity. Visually confirm that 4DM placed the heart volume of interest (VOI) (See 2, Figure 3) over the left ventricle (LV) or the entire myocardium, excluding any bone. Also confirm that the blood pool VOI (See 3, Figure 3) is placed in the LV outflow track at the base of the heart. If the VOI needs to be repositioned, left-click and hold within the VOI, dragging with your mouse accordingly. To adjust the size of the heart VOI, hover over the heart VOI until a red dot appears. Left click and hold the red dot to adjust the size. The blood pool VOI is not resizable.

In the Volumetric ROI Result table (See 4, Figure 3) the max Cardiac Metabolic Volume (CMV) and max Cardiac Metabolic Activity (CMA) values help you assess the uptake in myocardium. They provide an estimate of cardiac impact from tracer uptake in the heart at least 50% above the blood pool activity. Higher CMV and CMA values are believed to relate to higher amounts of amyloid in the heart. CMV and CMA values of 0 indicate a normal study. This review helps determine the semi-quantitative grade for the report.

Images

The Images screen is activated in the amyloid workflow when loading a reconstructed tomographic SPECT dataset. Review the images for cardiac uptake, extra cardiac activity, and regional differences of uptake (such that the brightness of the heart is inconsistent, indicating recent infarct) following the same process as for a SPECT MPI dataset. Uptake in the heart (See 1, Figure 4) brighter than that in the rib / bone (See 2, Figure 4), is an indication of Amyloidosis.

NOTE: Document your images review in the Qualitative Evaluation in the report.
Additionally, this visual review helps determine semi-quantitative grade in the report.

Volumetric Values	Description
Max	The max SUV value found per region.
Volume	The overall volume found in 3D ROI.
Threshold	Threshold is found by multiplying the meanblood pool SUV by 1.5.
CMV - Cardiac Metabolic Volume	Volume of activity above threshold within the Heart ROI in milliliters.
CMA - Cardiac Metabolic Activity	Activity in grams (g) calculated by multiplying your CMV value by the mean activity within the Heart VOI.

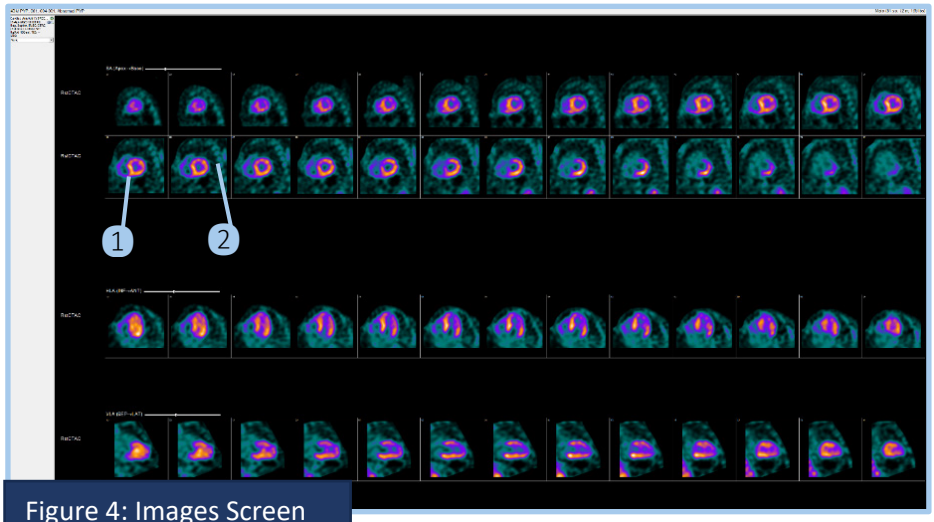

Figure 4: Images Screen

Reporting

4DM reporting has enhanced templates for Amyloidosis Rest and Amyloidosis Rest with CT available in 4DM version v2025 and higher. These are designed to decrease report turnaround times with less manual data entry. They follow ASNC, IAC and ACR reporting guidelines.

Figure 5: Select Reporting

To utilize these templates follow these steps:

1. Select reporting in your control panel (See 1, Figure 5).
2. The amyloidosis template will automatically open based on imaging protocol. (See Figure 6)
3. Review patient info, stress/EKG, and imaging info sections, which automatically populate based on the patient file.
4. In the Amyloidosis and CT Findings sections (See 1, Figure 6), use quick-select choices to efficiently populate text in your report. (See 2, Figure 6).
5. In the Technetium Uptake Section (See 3, Figure 6) the physician manually enters their qualitative evaluation of the patient, semi-quantitative grade and the quantitative heart/contralateral ratio that was calculated on the NM viewer.
6. Manually input any summary notes. (See 4, Figure 6)
7. Remember to save your work before closing 4DM. (See 2, Figure 5)

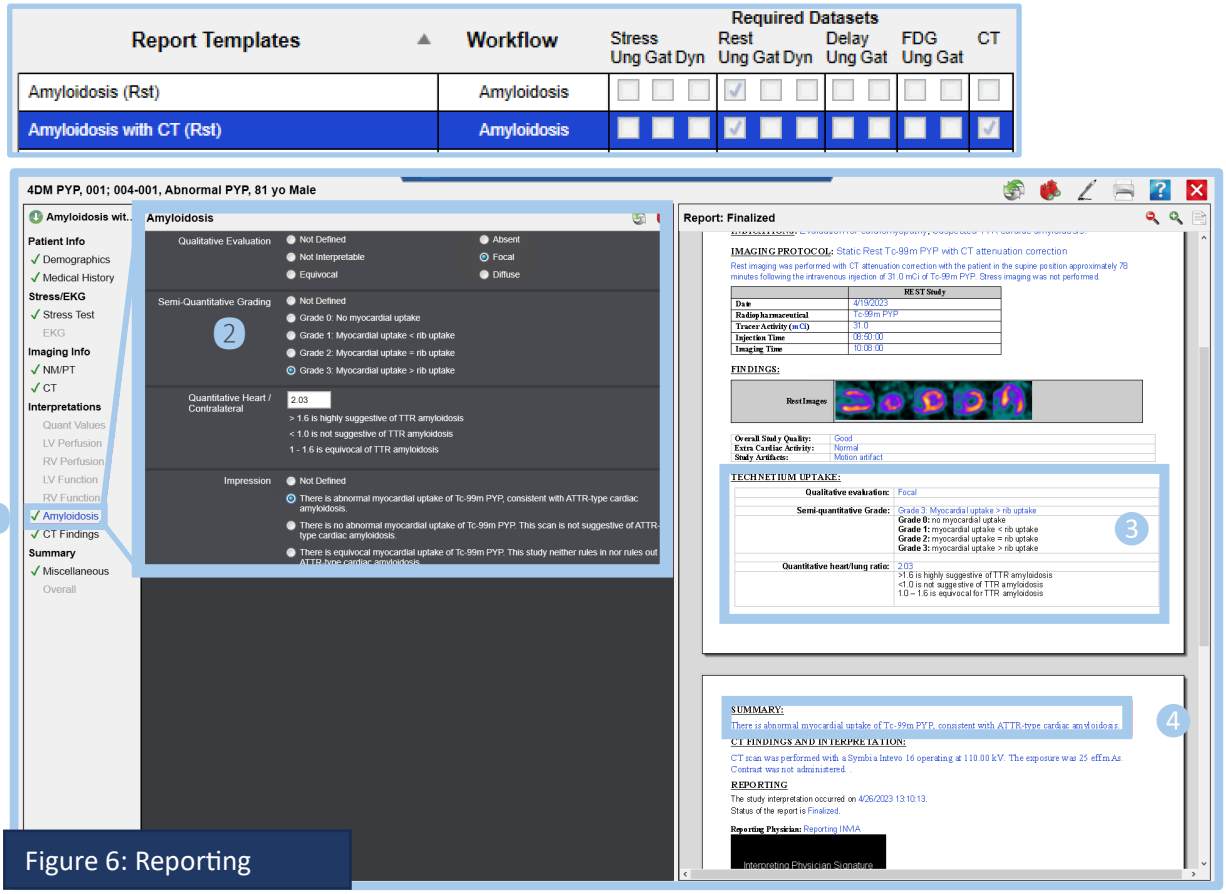

Figure 6: Reporting

HS_2025-49A