TRANSTHYRETIN AMYLOID CARDIOMYOPATHY (ATTR-CM) COMMUNICATION GUIDE

Patient Assessment at Each Visit

- **Eye issues**
  - Dark floater
  - Dry eyes
  - Glaucoma
  - Abnormal blood vessels
  - Pupillary abnormalities

- **Cardiovascular manifestations**
  - Dyspnea
  - Orthopnea
  - Paroxysmal nocturnal dyspnea (PND)
  - Abdominal/peripheral edema
  - Fatigue
  - Exercise intolerance
  - Dizziness/syncope
  - Palpitations/irregular heartbeats

- **Nephropathy**
  - Decline in renal function
  - Protein in urine

- **Gastrointestinal (GI)**
  - Nausea/vomiting
  - Diarrhea and/or constipation
  - GI bleeding
  - Pseudo-intestinal paralysis

- **Spinal stenosis**: Pain, tingling, or numbness in legs and/or buttocks

- **Peripheral sensorimotor**
  - Weakness, numbness, pain in hands/feet with progression to central portion of body
  - Walking/balance
    - ID safety issues/modifications needed (rugs, stairs, wheelchair accommodations)
  - Bladder problems

- **Activity levels**: general level, increased need for napping

- **Self-care**
  - Ability to carry out activities of daily living
  - Needed occupational therapy/assistive devices

- **Nutrition**
  - GI symptom management to improve poor intake, nutritional deficiencies, malabsorption
  - Swallowing difficulties
  - Heartburn
  - Early satiety
  - Sodium and fluid restriction
  - Daily weights
  - Unintentional weight loss
  - Daily caloric intake
    - Nutrition consult, regular follow-up

- **Mood**
  - Anxiety
  - Depression
  - Tools for screening

- **Altered sleep pattern factors**
  - Inadequate pain control
  - Sleep disordered breathing (SDB)
    - Nocturnal oxygen desaturation
  - Extra-cardiac factors
    - Macroglossia
    - Pharyngeal soft tissue thickening
    - Peripheral neuropathy
    - Dysautonomia

- **Medication reconciliation and adherence**
  - Financial constraints
  - Medication side effects
  - Access to medications
  - Medication management
Patient Education

Understanding health information and making related decisions is critical to a patient’s well-being. In addition to reading, writing, speaking, numeracy and critical analysis skills, other health literacy factors include experiences with health care systems, cultural and linguistic factors, format of materials, and how information is communicated.

Steps to help improve health literacy:
1. Keep information relevant
   a. Consider patient background, age, experience, and culture
2. Use plain language
3. Give small amounts of information; repeat it
   a. Use “teach back” method: ask a patient to tell you what you just told them
4. Confirm patient understanding
5. Explain in different ways as needed
6. Encourage questions
7. Use more than one teaching method
   a. Visual aids, 1:1 communication, digital or written materials
8. Consider potential barriers to learning, such as vision or hearing impairment

Clinic Visits and Care Team

Amyloid fibers may build up in cardiac tissue, nerves, and GI tract.

Primary care provider(s) (PCPs)
- May coordinate specialist visits and manage common symptoms such as hypotension, erectile dysfunction, early satiety

Cardiology (heart failure/rhythm specialists)
- Possible concentric left ventricular hypertrophy and cardiac dysfunction
- Screening and evaluations: echocardiogram, electrocardiogram, cardiac MRI, cardiac pyrophosphate (PYP) scan, measurement of cardiac biomarkers (i.e., troponin and natriuretic peptides)
- Diagnostic testing: cardiac biopsy or nuclear scintigraphy

Neurology
- Potential peripheral and/or autonomic neuropathy (i.e., carpal tunnel syndrome)
- Screening and evaluations: nerve conduction study, neuropathy impairment score (NIS), hypotension evaluation
- Diagnostic testing: nerve biopsy

Gastroenterology
- Possible chronic constipation, diarrhea, inadvertent weight loss, and/or appetite loss
- Screening and evaluations: motility studies, endoscopy or colonoscopy, measurement of iron levels
- Diagnostic testing: GI tract biopsy

Genetic counselors
- Definitive differentiation between wild type and hereditary transthyretin amyloidosis
- Help patients understand and adapt to medical, familial and psychological aspects of a hereditary condition
- Genetic testing: Sequencing TTR gene, typically completed by a blood or saliva sample

Other providers
- Deposits may less commonly affect the eyes, kidneys, leptomeninges

Pharmacy
- Help provide support with new medications and symptom relieving medications

Tips for Interprofessional Communication

Utilize best practices in care coordination
- Establish accountability and agree on responsibility
- Communicate/share knowledge
- Help with transitions of care
- Assess patient’s needs and goals
- Create a proactive care plan
- Monitor and follow-up, including responding to changes in patient’s needs
- Support patient’s self-management goals
- Link to community resources
- Work to align resources with patient and population needs

Establish a referral network in your area

Document all communication and actions in a timely manner adhering to facility-specific protocols and in accordance with HIPAA regulations
Resources for Patients and Families with Amyloidosis

Patient and family resources (national and local):
- amyloidosissupport.org
  + National and regional support group meetings
  + 24-hour hotline
  + Condition information; clinical trials
- oneamyloidosisvoice.org
  + Online community with a “social wall” to connect individuals
  + Amyloidosis resources and information
- arci.org (Amyloidosis Research Consortium)
  + Patient focused webinars
  + My Amyloidosis Pathfinder (treatment centers and clinical trials)
  + Support groups list
- amyloidosis.org (Amyloidosis Foundation)
  + Patient-focused webinars, brochures, videos
  + Amyloidosis fact sheets
  + Caregiver resources
  + Clinical trials information

Financial, emotional, other support:
- Financial Assistance: Nurse case managers affiliated with the pharmaceutical company may be assigned to a patient when the medication is prescribed, or patients can enroll directly
  + alnylamassist.com
  + akceaconnect.com
  + vyndalink.com
- Pharmaceutical company representatives will help navigate insurance coverage and benefit options
- Pharmaceutical companies also have emotional support programs

Companies will also provide regular check-ins with the patient and caregivers; may assist with transportation/lodging related to clinic visits/treatments

Care navigator resources:
- Local home health agencies: available patient services
- Social workers
- Community resources for patients
- Local faith communities: transportation, meals, respite care services

Professional resources:
- There are no current published national or international guidelines for the management and diagnosis of hereditary transthyretin amyloidosis. Starred (*) resources in the reference list may be of help in diagnosis and management.
- Amyloidosis Research Consortium (arci.org)
  + Education and resources for health care providers
- International Society of Amyloidosis (isaamyloidosis.org)
  + Premiere international society for amyloidosis researchers and health care professionals; biannual international conference
- A Guide for the Diagnosis and Management of Amyloidosis
  + Textbook from American Association of Heart Failure Nurses (AAHFN)
- Amyloidosis Clinical Resources App
  + Free smart phone app for health care providers to help with patient diagnosis/management
References:


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