Myeloma Academy

MyelomaUK

AL amyloidosis

Myeloma Nurse Guide

The Myeloma Nurse Guide Series has been developed to enhance nurses' knowledge, inform practice and support nurses in the delivery of high-quality treatment and care to people with myeloma and related conditions. The information has been reviewed by myeloma nurse and medical experts and should be used in conjunction with local and national policies, protocols and guidelines.

What is AL amyloidosis?

AL amyloidosis stands for amyloid light chain amyloidosis. In AL amyloidosis, amyloid fibrils, composed of abnormal light chain proteins, are produced by abnormal plasma cells in the bone marrow. These fibrils are deposited in tissues and organs of the body where they progressively accumulate and cause symptoms and disruption of organ function. The heart and kidneys are usually most affected, but patients often also have other organ/tissue involvement. Amyloid deposits in the heart can lead to progressive congestive cardiac failure and the risk of arrhythmias. The presence of cardiac amyloid is particularly important as it can significantly affect survival.

In the UK 500–600 new cases of AL amyloidosis are diagnosed each year. AL amyloidosis is a plasma cell disorder but as the plasma cells are not truly cancerous, it is not regarded as a cancer. It is however closely related to myeloma and is diagnosed in 10–20% of myeloma patients. Although treatment of AL amyloidosis is led by a haematologist, a multidisciplinary approach is required to manage disease complications and treatment side effects. There is currently no curative treatment available for AL amyloidosis, and like myeloma, relapse is common.

Amyloidosis is a complex disease and although AL amyloidosis is the most common form, other types exist. These include ATTR (transthyretin amyloidosis) of which some types are inherited, and AA amyloidosis which can be related to inflammatory conditions. These forms of amyloidosis are managed by different teams, not haematology.

Clinical features

Symptoms of AL amyloidosis are related to where amyloid fibrils are deposited in the body:

- **Heart:** Shortness of breath, palpitations, fatigue, dizziness, pulmonary and peripheral oedema
- **Kidneys:** Frothy urine, oedema of ankles, legs or abdomen, nephrotic syndrome
- **Gastrointestinal (GI) tract:** Reduced appetite, macroglossia (enlarged tongue, which may cause dry mouth and sleep apnoea), bloating, flatulence, diarrhoea/constipation, early satiety, enlarged liver or spleen, dysphagia, weight loss
- **Soft tissue and vascular system:** Bruising or bleeding, periorbital bruising, thrombosis, carpal tunnel syndrome
- **Skin and nails:** Lesions on face, trunk or limbs, waxy skin, weak nails
- **Peripheral nervous system:** Neuropathic pain, altered sensation, tingling
- **Autonomic nervous system:** Altered bowel habits, orthostatic hypotension, erectile dysfunction, sweating after eating

Appearance of any of these symptoms in a myeloma patient, at any point in their disease course, should raise the suspicion of AL amyloidosis.

Diagnosing and monitoring

Diagnosis is often delayed due to the rarity and range of symptoms that may occur. Patients with confirmed or suspected diagnosis of AL amyloidosis are often referred to a specialist unit, such as the National Amyloidosis Centre (NAC), for diagnostic and management advice. Assessment of organ function is key to evaluating treatment and managing symptoms. Monitoring patients' overall health is important to identify current and future supportive care needs.

Diagnostic and assessment features	Rationale
Tissue biopsy (from bone marrow, fat aspirate or affected organ), amyloid typing	To establish diagnosis of AL amyloidosis
Serum and urine immunofixation, sFLC, bone marrow biopsy, FISH, WBMRI, PET-CT	To detect abnormal plasma cells and monitor treatment response
Cardiac investigations: ECG, echocardiogram, NT-proBNP, troponin-T, cardiac MRI Renal and liver investigations: eGFR, 24-hour urine for proteinuria, alkaline phosphatase, albumin level Clotting profile SAP (serum amyloid protein) scan*	To assess the presence or extent of organ involvement To manage risk of complications To monitor treatment response
Vital signs, including lying and standing blood pressure Daily weight, fluid balance, nutritional intake Bowel function Neuropathic pain or numbness Skin changes, bruising, oedema Sleep pattern and presence of sleep apnoea	To pick up signs of AL amyloidosis To detect any changes in organ function and monitor overall health status

*Only available at the National Amyloidosis Centre

Treatment and management

Given the systemic nature of the disease, management of AL amyloidosis usually involves collaboration between specialist teams, such as haematology, nephrology, cardiology, gastroenterology and neurology. The aims of treatment are to: reduce abnormal plasma cells and their light chain production, reduce organ damage and improve function. Amyloid deposits may be slowly cleared by the body, but this can take many years. Providing supportive care to manage organ-specific complications is therefore crucial in optimising quality of life.

Treatment regimens are similar to those used in myeloma and may include steroids, bortezomib, cyclophosphamide, lenalidomide and daratumumab. Some patients may undergo autologous stem cell transplantation and, for selected patients in disease remission, organ transplants might be considered.

Management of cardiac amyloidosis is individualised and close working with the cardiology team is important. Treatments for cardiac symptoms include midodrine for orthostatic hypotension and diuretics for congestive cardiac failure. Calcium channel blockers are generally contraindicated in cardiac amyloidosis. Beta blockers and ACE inhibitors are poorly tolerated and their use should be carefully discussed with the cardiologists. Some patients may be suitable for an implantable cardiac device (such as a pacemaker or defibrillator) to manage the risk from arrhythmias.

Renal management can include diuretics, modifications to diet (such as a low-salt diet) and fluid restriction. Infusions of salt-poor albumin may help maintain intravascular volume in patients with nephrotic syndrome. End-stage renal failure is treated by dialysis.

Given the complex needs of this patient group, personalised and holistic care requires close liaison between the nursing team and other health professionals such as the dietitian, physiotherapist, psychologist and palliative care team.

Nursing management points

Assessment and monitoring

- Explain to patients the importance of regular monitoring and reporting any new or changing symptoms
- Assess for symptoms of neuropathy which patients may not report (e.g. erectile dysfunction, bowel changes)
- Educate patients to record their weight daily as required. Be aware that fluid retention may mask weight loss caused by malnutrition or malabsorption.
- If indicated, teach patients how to monitor their blood pressure and pulse at home. This might include doing their lying and standing blood pressure.
- Assess for signs of sleep apnoea, using a tool such as the STOP-Bang questionnaire academy.myeloma.org.uk/resources/stop-bang-questionnaire/

Treatment and management

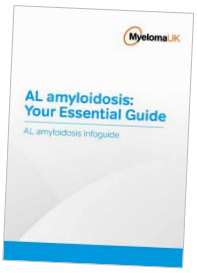
- Reinforce advice on any dietary restrictions in place e.g. low-salt, low-fibre diet
- Advise on strategies to help keep to oral fluid restrictions
- Artificial saliva may help relieve a dry mouth, particularly if the patient has macroglossia
- If required, refer patients to a dietitian to ensure adequate nutritional intake and liaise with GP regarding nutritional supplements
- Advise on management of constipation and diarrhoea
- Liaise with the medical team regarding blood pressure and weight changes, particularly low blood pressure and weight gain, especially if patient feels symptomatic
- Monitor response to prescribed analgesics for peripheral neuropathy, such as gabapentin, pregabalin and duloxetine
- Support stockings may be helpful for lower limb oedema
- Use of a wrist brace may help relieve symptoms of carpal tunnel syndrome
- Liaise as required with other teams, such as renal dialysis, cardiology and heart failure nurses, palliative care to promote personalised care planning
- Advise on resources available for support, noting that patients without a co-existing cancer diagnosis may not be able to access cancer support organisations

Self-care strategies for patients

- Diet and fluids
  - Eat little and often if appetite is poor or if easily full when eating
  - Reduce salt intake as advised. Foods which have a high salt content include processed foods, ready meals, crisps, bacon and canned meat.
  - If possible, eat energy-dense foods
  - Record your fluid intake and stick to any oral fluid restriction advised
- Skin care
  - Keep skin well moisturised, use emollient creams such as cocoa butter
  - Avoid tight fitting footwear and keep legs elevated when sitting
- Managing dizziness
  - Move slowly when rising from a sitting or lying position
  - Pass urine sitting down
- Information and support
  - Check AL amyloidosis resources from Myeloma UK. Consider joining a support group or discussion forum.
  - Ask about any financial support available
  - Keep a diary to note symptoms and have a record of consultations

For more detailed information on symptom management and treatment side effects, see Myeloma Nurse Guides on myelosuppression, myeloma kidney disease, gastrointestinal toxicities, fatigue, steroids, peripheral neuropathy, emotional and psychological support.

Patient information key points



- Provide written information to help patients understand their diagnosis
- Explain how to recognise and report symptoms and side effects
- Educate patients on ways to manage the effects of organ dysfunction

A video for AL amyloidosis patients
youtu.be/mBuG9NJ3IZ4

References

A list of key references is available on Myeloma Academy:
academy.myeloma.org.uk/myeloma-nurse-guide-references

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For further nurse guides and other educational resources on myeloma and related conditions:
academy.myeloma.org.uk

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Myeloma UK 22 Logie Mill, Beaverbank Business Park, Edinburgh EH7 4HG
0131 557 3332 myelomauk@myeloma.org.uk Registered Charity No: SC026116