

**Cardiff and Vale (C&V) UHB Corporate Medicines Management Group  
Shared Care Committee  
SHARED CARE**

**Drug: RILUZOLE                      Protocol number:    CV 39**

**Indication: AMYOTROPHIC LATERAL SCLEROSIS (ALS)**

**General Guidance**

This protocol sets out details for the shared care of patients taking **riluzole** and should be read in conjunction with Cardiff and Vale corporate Medicines Management Group (cMMG) Shared Care General Guidance. Good communication is required between the specialist, GP and patient. The arrangements for prescription and monitoring should be explained to the patient by the doctor(s) recommending and initiating treatment. **The doctor who prescribes the medication legally assumes responsibility for the drug and the consequences of its use.** The prescriber has a duty to keep themselves informed about the medicines they prescribe, their appropriateness, effectiveness and cost. They should also keep up to date with the relevant guidance on the use of the medicines and on the management of the patient's condition.

**Background**

Riluzole is indicated to extend life or the time to mechanical ventilation for patients with ALS. Riluzole therapy should only be initiated by specialist physicians with experience in the management of motor neurone disease (MND)

The drug has been evaluated and recommended in NICE Technology Appraisal 20, 2001, as being suitable for prescribing under Shared Care protocol arrangements. The umbrella term "Motor Neurone Disease" is used to describe three main sub-groups of the disease;

Amyotrophic Lateral Sclerosis (ALS) - mixed upper and lower motor neuron signs – this is the commonest form and the type for which riluzole is licensed. It is a progressive, fatal neurodegenerative disorder, characterised by both upper and lower motor neurone signs. Death usually results from ventilatory failure, resulting from progressive weakness and wasting of respiratory and bulbar muscles within approximately 3 years of the onset of symptoms.

Progressive Bulbar Palsy (PBP) - where the brunt of the disease at presentation falls upon the bulbar musculature

Progressive Muscular Atrophy (PMA) - lower motor neuron signs only

The guidance from NICE emphasises the use of riluzole in ALS, while noting that some clinicians have in certain cases prescribed in PMA. Pure upper motor neuron syndromes (e.g. primary lateral sclerosis, pseudobulbar palsy) are not licensed indications for riluzole.

Patient liaison may be facilitated either by visiting a hospital clinic or via the MND Care-Coordinators based at Rookwood Hospital (tel. 02920 313828)

Riluzole is the only drug currently licensed for the treatment of ALS. However symptomatic management, supportive and palliative care is also available for patients with ALS.

## **Epidemiology**

Incidence of ALS 2 per 100,000/year

Prevalence of ALS approximately 7/100,000

Average survival Progressive Bulbar Palsy form – 6 months to 3 years from symptom  
Average ALS – 2 -5 years from symptom onset

## **Dosage in adults**

Riluzole is normally prescribed as oral tablets 50mg twice daily No further dosage increment is required.

## **Responsibilities**

### **A. Consultant responsibilities**

1. When treatment is **initiated** send Shared Care request form with Shared Care protocol to GP
2. Initiate therapy following full discussion with the patient of benefits and risks and following consideration of baseline haematological and biochemical parameters: these include evaluation of baseline blood count, renal and liver function.
3. Counsel patients that treatment with riluzole is not expected to improve their existing symptoms from ALS
4. Advise patients to seek an urgent full blood count if a febrile illness develops. Counsel patients on how to recognise signs of neutropenia, for example, sore throat, fever infection, non-specific illness or liver toxicity (e.g. nausea, vomiting, abdominal discomfort and dark urine)
6. When a GP positive response to SC has been received and patient has been stabilised send a letter to GP “handing over” the Shared Care of the patient to the GP.
7. Respond to any request from GP to review the patient due to adverse effects of therapy.
8. Advise the GP on continuing or stopping riluzole therapy following review of the patient and associated drug therapy
9. Notify GP if patient is failing to attend for appropriate monitoring and advise GP on appropriate action.

## B. General practitioner responsibilities

1. Within one week of receipt return the completed Shared Care request form to indicate whether or not willing to undertake Shared Care.
2. Prescribe riluzole after 3 months as part of the shared care agreement.
3. Monitor the general health of the patient
4. Report adverse effects of therapy to the consultant and the Medicines and Health Care products Regulatory Agency (MHRA)
5. To act on advice provided by the Consultant if patient does not attend for appropriate monitoring.

## C. Patient responsibilities

1. Consent to treatment with riluzole
2. Attend regular appointments with specialist centre and GP.
3. Report any side effects to the specialist or GP whilst taking riluzole.

### Monitoring in secondary care

**Before treatment:** FBC, LFTS, electrolytes and creatinine.

**During Treatment:** LFTs monthly for 3 months Measurements may be required more frequently if transaminases start to rise.

### Monitoring by GP

FBC should be checked in patients with febrile illness/signs of neutropenia.

LFT at months 6, 9 and 12 months then annually thereafter

#### Stop riluzole if any of the following occurs: -

WBC	< 3.5x10 <sup>9</sup> /L * and/or
Neutrophils count	<1.5x 10 <sup>9</sup> /L*
Platelets	< 150 x 10 <sup>9</sup> /L*
	AST/ALT > 5 times the upper limit of normal range greater or equal to 225 IU/L

### Side effects

Gastrointestinal –nausea, vomiting and abdominal pain.

Increases in transaminases- elevation of liver enzymes most commonly occurs in the first 3 months after commencing therapy and may subsequently decline on treatment

Other side effects include asthenia, headache, pain, dizziness, tachycardia, somnolence and circumoral paraesthesia.

Neutropenia has been noted rarely- 3 cases have been recorded in 5000 patients who have received riluzole- check patient's white cell count if any febrile illness reported.

Interstitial lung disease (ILD) (uncommon) - perform chest radiography if symptoms such as dry cough or dyspnoea develop and stop if ILD is diagnosed ( e.g. bilateral diffuse lung opacities) In the majority of reported cases symptoms resolved after riluzole discontinuation and symptomatic treatment

Hepatitis has been reported rarely.

Once commenced it is very uncommon for riluzole to have to be stopped on grounds of side effects.

### **Contraindications:**

Hepatic disease or in patients with baseline transaminases greater than 3 times the upper limit of normal.

Hypersensitivity to riluzole or any excipients

Pregnancy and breast feeding

Renal impairment-avoid as no information available.

Acute porphyria

### **Interactions – Minor**

There have been no clinical studies to evaluate the interactions of riluzole with other medicines.

Enzyme inhibitors (e.g. caffeine, diclofenac, diazepam, imipramine, theophylline, amitriptyline and quinolones) could potentially decrease the rate of riluzole elimination.

Enzyme inducers (rifampicin and omeprazole) could increase the rate of riluzole elimination.

### **Special recommendations**

Patients should be warned about the potential for dizziness or vertigo and advised not to drive or operate machinery if these symptoms occur.

### **Contacts**

Patients with MND are initially under the care of a range of consultants (below). The majority within the remit of the Dept of Neurology at Cardiff and Vale are also seen by the MND Care-Coordinators and attend multidisciplinary regional clinics run by the South Wales MND Care Network at Rookwood Hospital and in Cwm Taf Health Board. **If you suspect an adverse reaction stop riluzole and contact the secretary**

**of the relevant consultant (as below) or the MND Care Co-ordinators (Rookwood Hospital 02920 313828):**

Department of Neurology Consultants:

Dr Andrea Lowman 029 20713769

Dr Ellie Marsh 029 20742835

Dr R Corkill 029 20748730

Dr. P. E. Smith 029 20742834

Dr K Hamandi 029 2072 5434

Dr. J. G. Llewelyn 029 2074 6441

Dr. N. Robertson 029 2074 5403

Dr TP Pickersgill 029 2074 5564

Dr. TAT Hughes 029 2074 2833

Dr K Dawson,029 20746441

Dr Mark Wardle 029 20744166

Prof AE Rosser via 029 20742835

Dr Valentina Thomassini 029 20742835

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