SHARED CARE GUIDELINES

Stiripentol in Children and Adolescents

It is vital for safe and appropriate patient care that there is a clear understanding of where clinical and prescribing responsibility rests between Consultants and General Practitioners (GPs). This guideline reinforces the basic premise that:

- When clinical and / or prescribing responsibility for a patient is transferred from hospital to GP, the GP should have full confidence to prescribe the necessary medicines. Therefore, it is essential that a transfer of care involving medicines that a GP would not normally be familiar with, should not take place without the "sharing of information with the individual GP and their mutual agreement to the transfer of care.

These are not rigid guidelines. On occasion, Consultants and GPs may agree to work outside of this guidance. As always, the doctor who prescribes the medication has the clinical responsibility for the drug and the consequences of its use.

Licensed indication:
Stiripentol is licensed for use in combination with clobazam and valproate as adjunctive therapy of refractory generalised tonic-clonic seizures in children with severe myoclonic epilepsy in infancy (Dravet Syndrome) whose seizures are not adequately controlled with clobazam and valproate. It should only be used under specialist supervision.
Please note it is Amber for this indication only.

Dosage and administration:

Child 3–18 years initially 10 mg/kg in 2–3 divided doses: titrate dose over minimum of 3 days to max. 50 mg/kg/day in 2–3 divided doses in combination with clobazam and valproate.
Any dose adjustment must be done by hospital consultant only.
It should not be taken with milk, dairy products, carbonated drinks, fruit juice or with food or drink that contains caffeine.

Cautions and contraindications: Stiripentol is contra-indicated in children with history of psychosis.
Adverse effects: nausea, vomiting; aggression, anorexia, ataxia, drowsiness, dystonia, hyperexcitability, hyperkinesia, hypotonia, irritability, sleep disorders, weight loss; neutropenia; *less commonly* fatigue, photosensitivity, rash, and urticaria.
Monitoring requirements:
Children treated with stiripentol should have full blood count and liver function tests prior to initiating treatment and every 6 months thereafter.

Action to be taken if abnormal results/adverse effects: In the event of any adverse effects the specialist clinic should write a management plan and this will be shared with primary care. If adverse events are detected in a primary care setting then the specialist should be informed as soon as possible.

Drug interactions: Stiripentol may interact with anticonvulsants, antidepressants, antipsychotics, MAOI, mefloquine, orlistat. Stiripentol should not be given with milk, dairy products, carbonated drinks, fruit juice or food or drinks which contain caffeine. For full information see the current edition of BNFc.

Specialist responsibilities:

1. Confirm the diagnosis of Dravet's syndrome or intractable epilepsy.
2. Communicate details of diagnosis to the GP and ensure patient carer is an informed recipient in therapy.
3. Ensure that patients/carers understand their treatment regimen and any monitoring or follow up that is required.
4. Issue a medicines for children information leaflet.
5. Ensure that the patient/parent/carer understands the nature, effect and potential side effects of the drug
6. Ensure baseline investigations are normal before commencing treatment.
7. Initiate treatment and prescribe for 3 months until patient is stabilised on their epilepsy regimen and the treatment is shown to be effective and well tolerated.
8. The decision to initiate stiripentol and all advice for dose titration will be carried out by the consultant paediatrician.
9. Arrange shared care with the GP once stabilized on medication.
10. Communicate with patient's community pharmacy that patient will shortly be requesting a supply from them.
11. Clinical and laboratory supervision of the patient by blood monitoring and routine clinic follow-up on a regular basis.
12. Send a letter/results notification to the GP after each clinic attendance ensuring current dose, most recent blood results and frequency of monitoring are stated.
13. Discontinue medication if ineffective or intolerable side effects.

GP responsibilities:

1. Ensure that the patient/parent/carer understands the nature, effect and potential side effects of the drug before prescribing it as part of the shared care program and contact the specialist for clarification where appropriate.
2. Monitor patient's overall health and well being
3. Report any adverse effects of medication to consultant.
4. Provide the patient with monthly repeat prescriptions of medication once the specialist has recommended continuation therapy. The patient should allow at least 48 hours for the prescription from the GP to be generated once they have agreed to take on prescribing.
5. Not to initiate or change the anticonvulsant medication without referral to the consultant.
6. Contact the specialist if they do not agree with the treatment recommendation, or if there is a perceived problem with compliance or concordance, or if they have any questions about the management plan.

Patient and parent/carer responsibilities:

1. Agree to request prescriptions from the GP in good time; obtain the first GP prescription within 2 weeks of being informed that shared care will be in operation.
2. Report any concerns or adverse effects to the GP, paediatrician or pharmacist.
3. Ensure they have a clear understanding of their treatment.
4. Report any changes in disease symptoms to GP and/or specialist
5. Alert GP and/or specialist of any changes of circumstance which could affect management of disease e.g. plans for pregnancy
6. Take/ administer the medication as prescribed.
7. Undertake any monitoring as requested by the GP and/or specialist

Secondary care review: Patients will be reviewed at regular intervals, initially 6 to 8 weeks after starting medication. Children maintained on effective medication will be seen at least every 6 months and more frequently if needed, or if requested to review by the GP.

Availability: (BNFC, March 2016)
Capsules, stiripentol 250 mg (pink), net price 60-cap pack = £284.00; 500 mg (white), 60-cap pack = £493.00
Powder, stiripentol 250 mg, net price 60-sachet pack = £284.00; 500 mg, 60-sachet pack = £493.00.

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