Pharmacological management of Postural Orthostatic Tachycardia Syndrome in adults

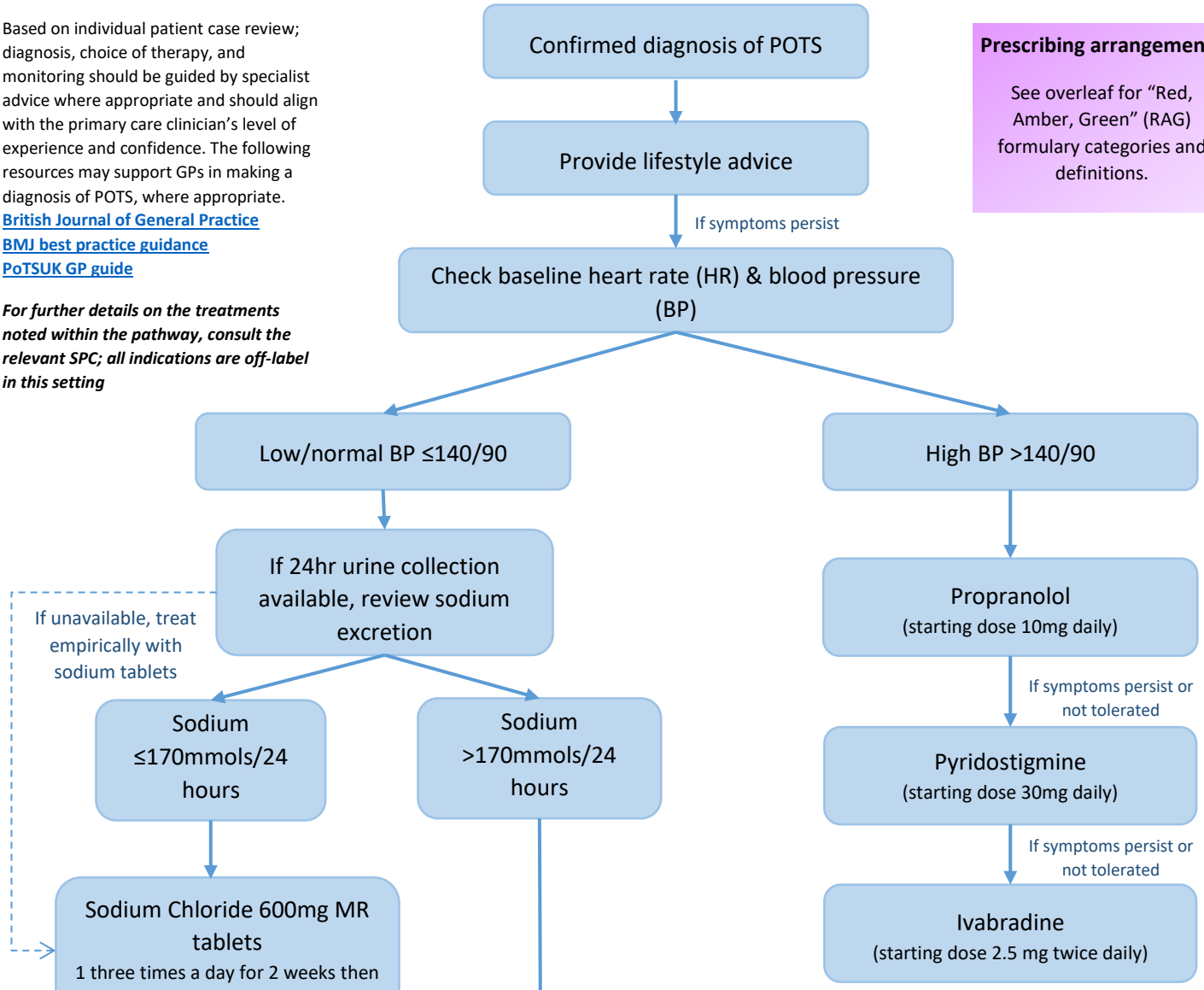
Based on individual patient case review; diagnosis, choice of therapy, and monitoring should be guided by specialist advice where appropriate and should align with the primary care clinician's level of experience and confidence. The following resources may support GPs in making a diagnosis of POTS, where appropriate.

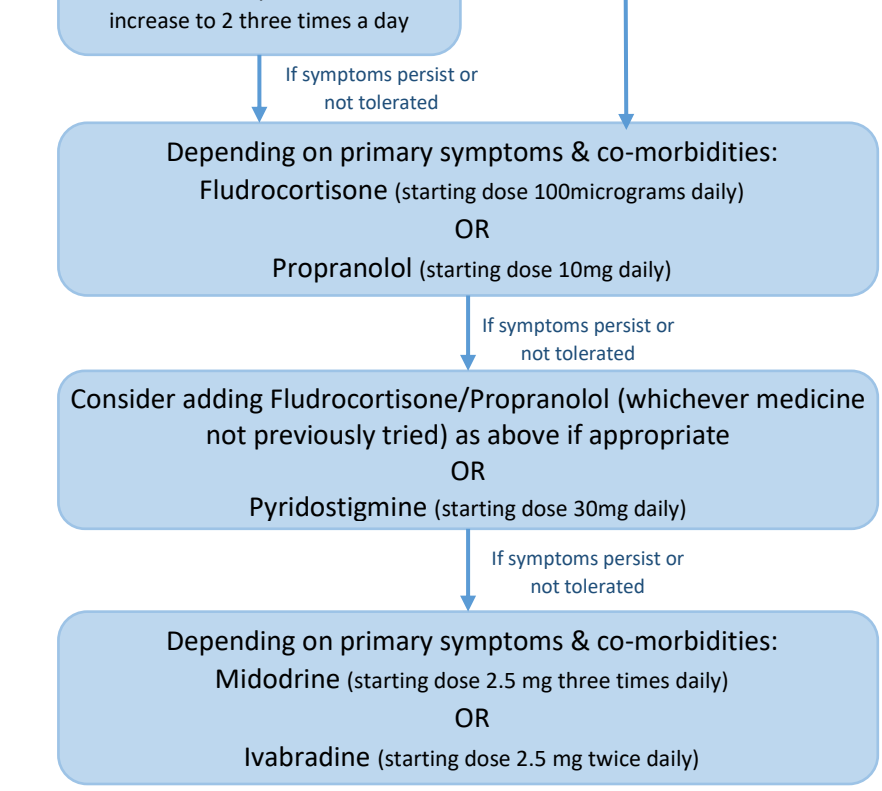
British Journal of General Practice
BMJ best practice guidance
PoTSUK GP guide

For further details on the treatments noted within the pathway, consult the relevant SPC; all indications are off-label in this setting

Prescribing arrangements

See overleaf for "Red, Amber, Green" (RAG) formulary categories and definitions.





Titration and Deprescribing

All medicines should be gradually titrated up over 6 weeks until maximum benefit is achieved – see page 2 for details on titration .

If there is no benefit after 6 weeks, the medicine should be stopped, and next line of therapy trialled.
If there is some benefit, but patient is still symptomatic, add in the next line of therapy.

When deprescribing, higher doses of propranolol and fludrocortisone may require weaning (see table on pages 2 & 3), but all other medications can be stopped without weaning and without withdrawal.

South East London Integrated Medicines Optimisation Committee (SEL IMOC). A partnership between NHS organisations in South East London Integrated Care System: NHS South East London (covering the boroughs of Bexley/Bromley/Greenwich/ Lambeth/Lewisham and Southwark) and GSTFT/KCH /SLaM/ Oxleas NHS Foundation Trusts and Lewisham & Greenwich NHS Trust
Approval date: October 2025 **Review date:** October 2027 (or sooner if evidence or practice changes)

LIFESTYLE ADVICE

The mainstay of therapy for POTS is lifestyle changes. These revolve around increased fluid and salt intake to increase blood volume, the use of compression clothing to support the circulation and building up exercise in a slow and gradual manner. On the list of blood tests we send out, there is a 24-hour urine collection for sodium and volume which provides us with an understanding about how well-hydrated patients are. Ideally, we want to see more than 2.5 litres of urine. If the volume is lower, this suggests that the patient should drink more fluid. If the sodium levels are lower than 170 mmol over 24 hours, this suggests that further increases in salt intake can be considered. Compression clothing can be formally prescribed and there are further details on the POTS-UK website, but often sports compression leggings will suffice. Patients should notice some improvement within a few weeks of implementing lifestyle changes.

POTS MEDICATION

A number of medications are recommended for treating POTS symptoms, and although use in POTS is off-label, these medications have been used for many years in this setting. Further information can be found about the medications used on POTS-UK website. **Medicines in this pathway are either Amber 1 (initiation in primary care on advice of the specialist) or Amber 3 (initiation/continuation by specialist until patient stable (min. 3 months prescribing), then transfer to primary care under shared care. See table below.**

The process for starting the medications will depend on the patient but usually one medication is started at the smallest dose and increased as per the instructions on the clinic letter. If the medication is increased to the maximum dose and produces no benefit, then it can be stopped. If it causes side-effects, it should be stopped. If it provides benefit, the optimal dose can be found and if symptoms warrant, the next medication can be added in. Some of the medications will be contraindicated in certain patients and some will be difficult because the heart rate becomes too slow or blood pressure too high. Each medication has individual instructions with regard to that aspect. The idea is to start one medication and assess its effects and add in the next medication as needed. All of the POTS medications can be used together if needed.

MONITORING & FURTHER SUPPLY

Patients will be encouraged to self-monitor their heart rate (HR) & blood pressure (BP) during initiation and once stabilised. They will report these to the arrhythmia nurses at regular intervals. Once benefit has been confirmed at 6 weeks and the patient is on a stable dose, the arrhythmia nurses will request that the GP takes over prescribing. For Amber 3 medicines, a further prescription will be issued at this point to make sure the patient has 3 months from the hospital, and the relevant paperwork will be sent to the GP.

CONTACTS

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Drug	Starting dose	Titration instructions	GP Monitoring	Additional comments
Sodium Chloride MR tablets (Slow Sodium) AMB 1	600mg three times a day	After 2 weeks, the dose can be increased to 1200mg three times daily This may be increased to 1800mg three times daily if needed.	Yearly BP & HR Yearly Urea & Electrolytes (U&E's) & Liver function tests (LFTs)	Alternatives: Patients can weigh out 6-10 g of salt at the beginning of the day and try to gradually add that to food during the day. Electrolyte tablets (e.g. Vitassium Capsules) are available to purchase online have a similar salt concentration and therefore the dose regime is very similar.
Fludrocortisone tablets AMB 1	100 micrograms daily	After 2 weeks, if potassium is normal, the dose can then be increased to 200 micrograms daily	Potassium 3 monthly Yearly BP & HR Yearly U&E's & LFTs	At the small doses prescribed, it does not have significant effects on the immune system nor does it have significant steroidal effects When stopping treatment, if taken for over 3 weeks, the dose will need to be gradually tapered down to avoid Adrenal crisis.

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Drug	Starting dose	Titration instructions	GP Monitoring	Additional comments
Propranolol tablets AMB 1	10mg daily	After 1 week increase to 10mg twice daily, then increase to 10mg three times a day and gradually add 10mg to each dose point until the maximum benefit is achieved. The maximum dose of propranolol is 320 mg daily in total but it is unlikely patients will get close to this dose.	Yearly BP & HR Yearly U&E's & LFTs	Contraindicated in asthma. When stopping treatment, doses over 160mg daily will need to be gradually tapered down to avoid withdrawal symptoms
Pyridostigmine tablets AMB 3 SEL formulary recommendation	30mg daily	After 1 week increase to 30mg twice daily and then to 30mg three times daily. The dose can subsequently be gradually increased all the way to 60mg three times daily.	Yearly BP & HR Yearly U&E's & LFTs	It is contraindicated in asthma and therefore should not be used in those patients. The major side effect is gastrointestinal disturbance which settles very easily by stopping the medication. • Transfer of Prescribing Responsibility
Ivabradine tablets AMB 3 SEL formulary recommendation	2.5mg twice daily	After 2 weeks increase to 2.5mg three times daily. Then add 2.5mg to each dose point every 2 weeks, thus increasing to 5/2.5/2.5, then to 5/5/2.5 and then to 5mg three times daily. The recommended maximum dose is 15mg daily - **Doses in excess of this are not recommended.**	Yearly BP, HR, pulse rhythm check Yearly U&E's & LFTs	MHRA Advice for the use of ivabradine in symptomatic angina also applies for POTS patients If HR falls persistently below 50 bpm (at rest) and/or the patient experiences symptoms related to bradycardia such as dizziness, fatigue or hypotension – This will require a dose reduction or cessation of therapy. If patient develops AF, ivabradine should be stopped. Ivabradine is a known teratogen – if prescribed in a woman of childbearing age ensure risks and benefits are discussed including the need for highly effective contraception. • Transfer of Prescribing Responsibility
Midodrine tablets AMB 3 SEL formulary recommendation	2.5mg three times daily	After 2 weeks increase to 5mg three times daily and then to 7.5mg three times daily and then to 10mg three times daily. The normal recommended maximum dose is 30mg daily - **Doses in excess of 40mg daily are not recommended.**	Yearly BP & HR Yearly U&E's & LFTs	Patients must self-monitor their HR and BP and stop the medication if it becomes high. Doses should not be taken within 4-5 hours of bedtime due to risk of supine hypertension. If patients do need to lie down because of fatigue but they are within 4-5 hours of a dose of midodrine, they should be propped up at 45 degrees and check their BP regularly. • Transfer of Prescribing Responsibility