European Porphyria Initiative

www.porphyria-europe.org

RECOMMENDATIONS FOR THE USE OF DRUGS IN THE ACUTE PORPHYRIAS (AIP, HCP, VP)

Drugs and provocation of acute attacks

This information provides guidance based on the available evidence on the time of publication (for the latest information see www.porphyria-europe.org).

However this evidence is not always complete, which may lead to some degree of uncertainty. It must be made clear that the prescription of drugs to a patient with acute porphyria is entirely at the responsibility of the physician in charge.European Porphyria Initiative members accept no responsibility for any error, difference of opinion or any adverse consequences arising from or occasioned by the use of information published on this website.

General principles on prescribing

When prescribing in acute porphyria, the benefit from using the drug of choice should always be assessed against the risk of provoking an acute attack and the likely consequences of not using it. A drug should never be withheld if it is judged to be essential for optimum treatment. Individuals may be encountered who have used an unsafe drug without adverse effects, even for many years, before being found – usually through family screening - to have latent porphyria. After discussion of the risk, such individuals often wish to continue on the drug and this should be allowed. However, if an additional drug needs to be prescribed, the possible risk of interaction provoking an acute attack needs to be carefully considered and a change to a safer combination may be required. Individuals who have inherited one of the acute porphyrias vary in their susceptibility to drug-induced acute attacks. It is known that many such individuals tolerate drugs known to be unsafe without adverse reaction. At present there is no way to identify in advance inter-individual variation in the response to drugs. Probable lower risk groups are those aged over 40 years who have never had symptoms, particularly men, and who have normal PBG excretion. Acute attacks are very rare in prepubertal children but the usual practice is to prescribe as for young adults. All individuals starting therapy should be advised to report any adverse reaction (eg abdominal pain) immediately. For all except safe/very low risk drugs, facilities should be available for prompt diagnosis and treatment of an acute attack, including 24 hour access to a supply of human hemin. For potentially unsafe drugs, monitoring urinary PBG excretion should be done in collaboration with a specialist porphyria centre to detect any drug-provoked increase.

Selecting a drug

Many drugs are not listed because their status is uncertain, often because there is insufficient experience of their use in acute porphyria – this applies particularly to newly introduced drugs

A list of potentially unsafe drugs is available on the European Porphyria Initiative website

www.porphyria-europe.org.

Drugs considered safe for use in the acute porphyrias

List A contains drugs in alphabetical order. In list B the same drugs are grouped by clinical indication.
If the drug is on list A and B it is considered safe.
If the drug is not on either lists:

- select an alternative from the recommended list
- consult the EPI website *www.porphyria-europe.org*
- consult the porphyria drug database *www.drugs-porphyria.com*

LIST A

Acetylcysteine
Aciclovir
Adrenaline
Alfentanil
Alimemazine
Allopurinol
Alpha tocopheryl
Alteplase
Aluminium salts
Amantadine
Amiloride
Aminoglycosides
Amphotericin
Ascorbic acid
Aspirin
Atovaquone
Atropine
Azithromycin
Barium sulphate
Beclometasone
Bendroflumethiazide
Beta blockers
Bezafibrate
Bisacodyl
Bismuth
Bisphosphonates
Bumetanide
Bupivacaine
Buprenorphine
Buserelin
Calcitonin
Calcium carbonate
Candesartan
Carbimazole
Chloral hydrate
Chloroquine
Chlorphenamine
Chlorpromazine
Ciclosporin
Cisplatin
Clobazam
Clofibrate
Clomifene
Clonazepam
Co-amoxiclav

Co-codamol
Codeine phosphate
Co-dydramol
Colchicine
Colestipol
Colestyramine
Corticosteroids
Corticotrophin
Cyclopenthiazide
Cyclopropane
Dalteparin
Dantron
Desferrioxamine
Desloratidine
Dexamethasone
Dextran
Dextromethorphan
Dextromoramide
Dextrose
Diamorphine
Diazoxide
Dicycloverine
Diflunisal
Digoxin
Dihydrocodeine
Dimercaprol
Dimeticone
Diphenhydramine
Diphenoxylate
Dipyridamole
Distigmine
Dobutamine
Domperidone
Dopamine
Doxorubicin
Droperidol
Enalapril
Enoxaparin
Epinephrine
Epoetin
Ethambutol
Ether
Famciclovir
Fenbufen
Fenofibrate
Fentanyl
Flucloxacillin
Flucytosine
Flumazenil
Fluoxetine
Fluphenazine
Flurbiprofen
Fructose
FSH
Furosemide
Gabapentin
Ganciclovir
Gemfibrozil
Glipizide
Glucagon
Glucose
Glycopyrronium
Gonadorelin
Goserelin
GTN
Guanethidine
Haloperidol
Heparin
Hetastarch
Hydrochlorothiazide
Hydrocortisone
Ibuprofen
Immunoglobulins
Indometacin
Insulin
Iron
Isoflurane
Ispaghula
Ketoprofen
Ketotifen
Lactulose
Leuprorelin
Levetiracetam
Levomepromazine
Levothyroxine
LHRH
Lidocaine
Lisinopril
Lithium
Loperamide
Loratadine
Lorazepam
Magnesium sulphate
Mefloquine
Melphalan
Mesalazine
Metformin
Methadone
Methylphenidate
Methylprednisolone
Mianserin
Midazolam
Morphine
Muscle relaxants non-depolarising
Naftidrofuryl
Nalbuphine
Naloxone
Naproxen
Neostigmine
Nitrous oxide
Octreotide
Olanzepine
Omeprazole
Ondansetron
Oxybuprocaine
Oxytocin
Paracetamol
Paraldehyde
Penicillamine
Penicillins
Pentamidine
Pethidine
Phentolamine
Phytomenadione
Pipotiazine
Prazosin
Prednisolone
Prilocaine
Primaquine
Procainamide
Procaine
Prochlorperazine
Proguanil
Promazine
Promethazine
Propantheline
Propofol
Propylthiouracil
Proxymetacaine
Pseudoephedrine
Pyridoxine
Pyrimethamine
Quinine
Resorcinol
Rosuvastatin
Salbutamol
Senna
Sodium acid phosph
Sodium bicarbonate
Sodium fusidate
Sorbitol
Streptokinase
Streptomycin
Sucralfate
Sulindac
Suxamethonium
Temazepam
Tetracaine
Thiamine
Tiaprofenic acid
Tinzaparin
Tranexamic acid
Triamterene
Trifluoperazine
Urokinase
Vaccines
Valaciclovir
Vancomycin
Vigabactrin
Vitamins
Warfarin
Zalcitabine
Zinc preparations
Zopiclone

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LIST B

Analgesics	Alfentanil
	Aspirin
	Buprenorphine
	Co-codamol
	Codeine phosphate
	Co-dydramol
	Dextromethorphan
	Dextromoramide
	Diamorphine
	Diflunisal
	Dihydrocodeine
	Fenbufen
	Fentanyl
	Flurbiprofen
	Ibuprofen
	Indometacin
	Ketoprofen
	Methadone
	Morphine
	Nalbuphine
	Naproxen
	Paracetamol
	Pethidine
	Sulindac
	Tiaprofenic acid
Antibacterial agents	Aminoglycosides
	Azithromycin
	Co-amoxiclav
	Ethambutol
	Flucloxacillin
	Penicillins
	Pentamidine
	Sodium fusidate
	Streptomycin
	Vancomycin
Anticonvulsants	Clobazam
	Clonazepam
	Gabapentin
	Levetiracetam
	Vigabactrin
Antidepressants	Fluoxetine
	Mianserin
Anti-emetics	Domperidone
	Ondansetron
	Prochlorperazine
	Promazine
Antihistamines	Alimemazine
	Chlorphenamine
	Desloratidine
	Diphenhydramine
	Ketotifen
	Loratadine
	Promethazine
Antipsychotics	Chlorpromazine
	Fluphenazine
	Haloperidol
	Levomepromazine
	Olanzepine
	Pipotiazine
	Trifluoperazine
Antivirals.antifungals	Aciclovir
	Amphotericin
	Famciclovir
	Flucytosine
	Ganciclovir
	Valaciclovir
	Zalcitabine
Cardiovascular agents	Alteplase
	Amiloride
	Bendroflumethiazide
	Beta blockers
	Bumetanide
	Candesartan
	Cyclopenthiazide
	Dalteparin
	Diazoxide
	Digoxin
	Dipyridamole
	Dobutamine
	Dopamine
	Enalapril
	Enoxaparin
	Furosemide
	Guanethidine
	Heparin
	Hydrochlorothiazide
	Lisinopril
	Naftidrofuryl
	Prazosin
	Procainamide
	Streptokinase
	Tinzaparin
	Triamterene
	Urokinase
Drugs used in anaesthesia	Adrenaline
	Atropine
	Cyclopropane
	Ether
	Isoflurane

	Muscle relaxants non-depolarising
	Neostigmine
	Nitrous oxide
	Phentolamine
	Propofol
	Suxamethonium
Immunisations	Immunoglobulins
	Vaccines
Lipid lowering agents	Bezafibrate
	Clofibrate
	Colestipol
	Colestyramine
	Fenofibrate
	Gemfibrozil
	Rosuvastatin
Local anaesthetics	Bupivacaine
	Lidocaine
	Prilocaine
	Procaine
	Tetracaine
Malaria prophylaxis	Atovaquone
	Chloroquine
	Mefloquine
	Proguanil
Miscellaneous	Acetylcysteine
	Allopurinol
	Alpha tocopheryl
	Aluminium salts
	Amantadine
	Ascorbic acid
	Barium sulphate
	Beclometasone
	Bisacodyl
	Bismuth
	Bisphosphonates
	Buserelin
	Calcitonin
	Calcium carbonate
	Carbimazole
	Chloral hydrate
	Ciclosporin
	Cisplatin
	Clomifene
	Colchicine
	Corticosteroids
	Corticotrophin
	Dantron
	Desferrioxamine
	Dexamethasone
	Dextran
	Dextrose
	Dicycloverine
	Dimercaprol
	Dimeticone
	Diphenoxylate
	Distigmine
	Doxorubicin
	Droperidol
	Epinephrine
	Epoetin
	Flumazenil
	Fructose
	FSH
	Glipizide
	Glucagon
	Glucose
	Glycopyrronium
	Gonadorelin
	Goserelin
	GTN
	Hetastarch
	Hydrocortisone
	Insulin
	Iron
	Ispaghula
	Lactulose
	Leuprorelin
	Levothyroxine
	LHRH
	Lithium
	Loperamide
	Lorazepam
	Magnesium sulphate
	Melphalan
	Mesalazine
	Metformin
	Methylphenidate
	Methylprednisolone
	Midazolam
	Naloxone
	Octreotide
	Omeprazole
	Oxybuprocaine
	Oxytocin
	Paraldehyde
	Penicillamine
	Phytomenadione
	Prednisolone
	Primaquine
	Propantheline
	Propylthiouracil
	Proxymetacaine
	Pseudoephedrine
	Pyridoxine
	Pyrimethamine
	Quinine
	Resorcinol
	Salbutamol
	Senna
	Sodium acid phosph
	Sodium bicarbonate
	Sorbitol
	Sucralfate
	Temazepam
	Thiamine
	Tranexamic acid
	Vitamins
	Warfarin
	Zinc preparations
	Zopiclone