

UKINETS bitesize guidance Management of Refractory Carcinoid Syndrome

Carcinoid syndrome is characterized by cutaneous flushing, gut hypermobility with diarrhoea and bronchospasm. Refractory carcinoid syndrome occurs when symptoms are no longer controlled by standard doses of somatostatin analogues (SSAs) (Octreotide LAR 30mg i.m. or Lanreotide autogel 120mg s.c. every 28 days).

Evaluate for tachyphylaxis and absorption issues such as fibrosis at injection site

Control of symptoms

Tumour control

High tumour burden in liver

Yes

No

Dose escalation of SSA

Increase frequency of long acting SSA to every 3 weeks or 2 weeks (1,2)
Continuous subcutaneous infusion of octreotide
Short acting octreotide s.c. can be used in addition for intermittently increased symptoms
Could consider antihistamines (cyproheptadine or chlorphenamine) +/- oral steroids (i.v. if carcinoid crisis)

Telotristat ethyl 250mg t.i.d. An oral tryptophan hydroxylase inhibitor. Approved for treatment of diarrhoea associated with CS in combination with SSA in patients insufficiently controlled by SSA. Not for routine commissioning in the NHS (7,8) depending on local access

Liver-directed therapies and tumour debulking to reduce the volume of tumour producing hormones (9,10)

Embolisation, resection, RFA, Liver transplantation (in very selected cases, see UKINETS bitesize guidance)*# following MDT discussion
*Patients with carcinoid syndrome are at risk for developing carcinoid crisis during surgery or other types of intervention (See perioperative management of patients with neuroendocrine tumours, UKINETS bitesize guidance).
#Carcinoid heart disease must be managed prior to surgery

SSA should be continued throughout treatment lines

Dose escalation of SSA

Peptide receptor radionuclide therapy (3)*

Everolimus and SSA (off label) (4)

Interferon alpha (3-5 million IU sc three times weekly) in combination with SSA
Associated with significant toxicity (5)

Pasireotide LAR may be considered off label in very select patients when other treatments fail or are not feasible (6)

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References

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