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AN OVERVIEW OF CARCINOID SYNDROME
CARCINOID SYNDROME (CS): WHAT IS IT?

• Paraneoplastic syndrome
• Peptides release (i.e. serotonin, histamine, kallikrein)
• Neuroendocrine tumours
• Liver metastases
CLINICAL FEATURES

GI NETs (67%)
- 38% midgut (mostly ileal NETs)
- Multifocal: 30% of cases
- Age: 60-70
- Typical CS (95%)

THORACIC NETS (25%)
- Mostly: typical bronchial carcinoid
- Age: 50’s
- Atypical CS (5%)

CS, carcinoid syndrome; GI, gastrointestinal; NET, neuroendocrine tumour
• **TYPICAL** CARCINOID SYNDROME
• **ATYPICAL** CARCINOID SYNDROME
TYPICAL CARCINOID SYNDROME

- FLUSHING: Face/neck/superior thorax, dry skin
- DIARRHOEA: Chronic, watery, no improvement if fasting, electrolitic alterations
- ABDOMINAL PAIN: Constant/intermittent, dull/cramp-like pain, no improvement after defecation
ATYPICAL CARCINOID SYNDROME

- Flushing
- Bronchospasm / Hypotension
- Eyelid Oedema
- Prolonged
Carcinoid cardiopathy

- 10-20% at NET diagnosis
- Cardiac right valve fibrosis
- Right cardiac failure
- 50% death

NET, neuroendocrine tumour
CARCINOID SYNDROME: DIAGNOSIS

NET, neuroendocrine tumours; NSAIDs, non-steroid anti-inflammatory drug; 5-Fu, Fluorouracil; 5-HIAA, 5-hydroxyindoleacetic acid; 11115-HTP, 5-Hydroxytryptophan

Figure reproduced from: Harrison’s Principles of Internal Medicine, 19th Edition. Accessed via www.accessmedicine.com
CARCINOID CRISIS

- Life Threatening
- Oncological Emergency

- Anaesthesia
- Interventional procedures
- Drugs
- Alcohol, foods, physical/psychological stress, infections

- Hypotension (rarely hypertension), tachycardia, dyspnoea & central nervous system symptoms
THERAPEUTIC OPTIONS

• LDT
• Surgery
• IFN
• Somatostatin analogs
• PRRT
• Telotristat ethyl*

*Telotristat ethyl is indicated for the treatment of CS diarrhea in combination with SSA therapy in adults inadequately controlled by SSA therapy

IFN, interferon LDT, liver directed therapies; PRRT, peptide receptor radionuclide therapy
CONCLUSIONS

• Carcinoid syndrome is present in around 20% of patients with NETs
• Linked to poorer prognosis
• Number of therapeutic options currently available with more options under study:
  – Increase SSA doses
  – Everolimus (if available)
  – PRRT
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Email antoine.lacombe@cor2ed.com
Dr. Antoine Lacombe
Pharm D, MBA
Phone: +41 79 529 42 79
antoine.lacombe@cor2ed.com

Dr. Froukje Sosef
MD
Phone: +31 6 2324 3636
froukje.sosef@cor2ed.com