

# Carcinoid Tumours and Carcinoid Syndrome - What we know -What we don't know

Dr Anthony Chambers MS FRACS  
Endocrine Surgeon & Surgical Oncologist  
St. Vincent's Hospital, Sydney

Unicorn Foundation Patient Support Meeting  
North Shore Private Hospital  
St Leonards, NSW 14<sup>th</sup> April 2010





UNIVERSITY OF  
CALGARY









Dr Janice Pasieka



# Neuroendocrine Tumour Clinic

## University of Calgary

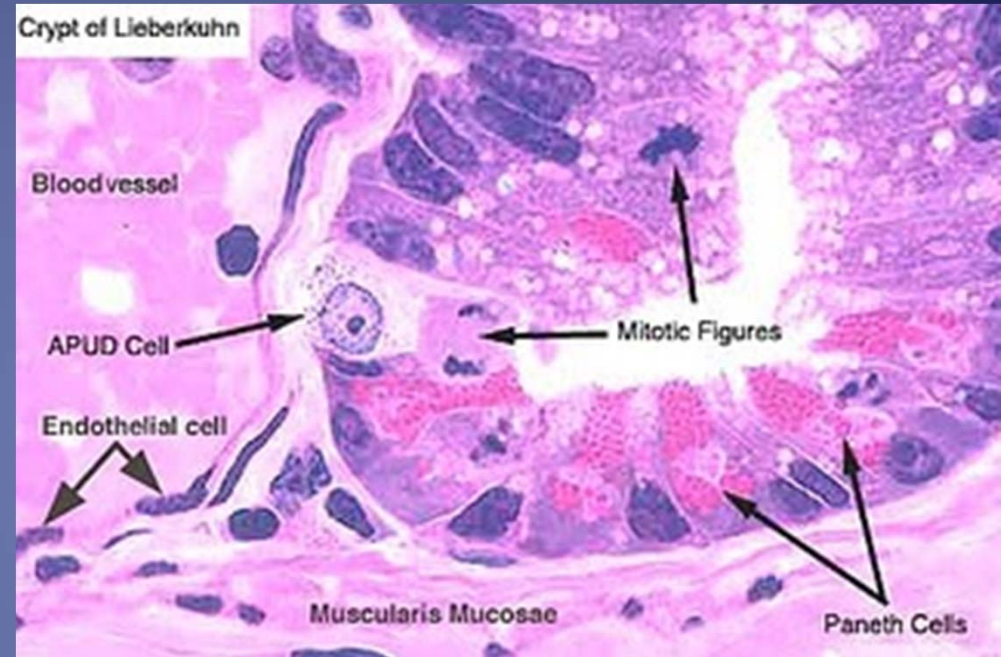
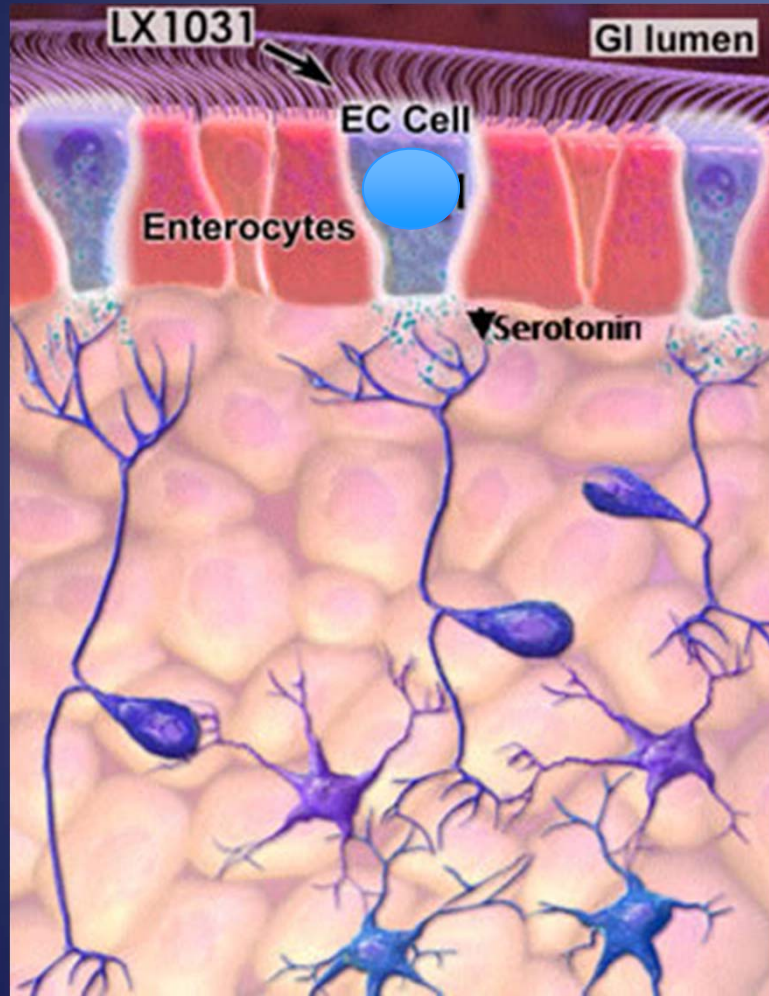
- 15 years
- Multidisciplinary
  - Endocrinologist – Dr Otto Rorstad
  - Endocrine Surgeon – Dr Janice Pasieka
  - Hepatobiliary Surgeon – Dr Elijah Dixon
  - Nuclear Medicine Physician – Dr Ernst Postema
  - Clinical Geneticist – Dr Michael Innes
- Referral center for Western Canada

# Gastrointestinal NET

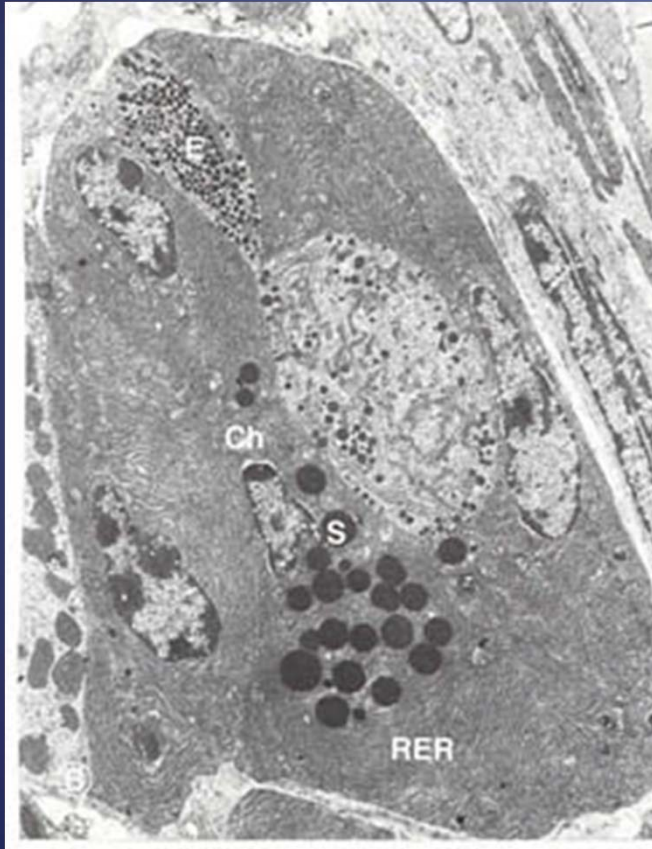
- Tumours derived from Enterochromaffin Cells (EC cells)
- Most common sites
  - Small intestine
  - Appendix
  - Colon & Rectum



# Enterochromaffin (EC) cells

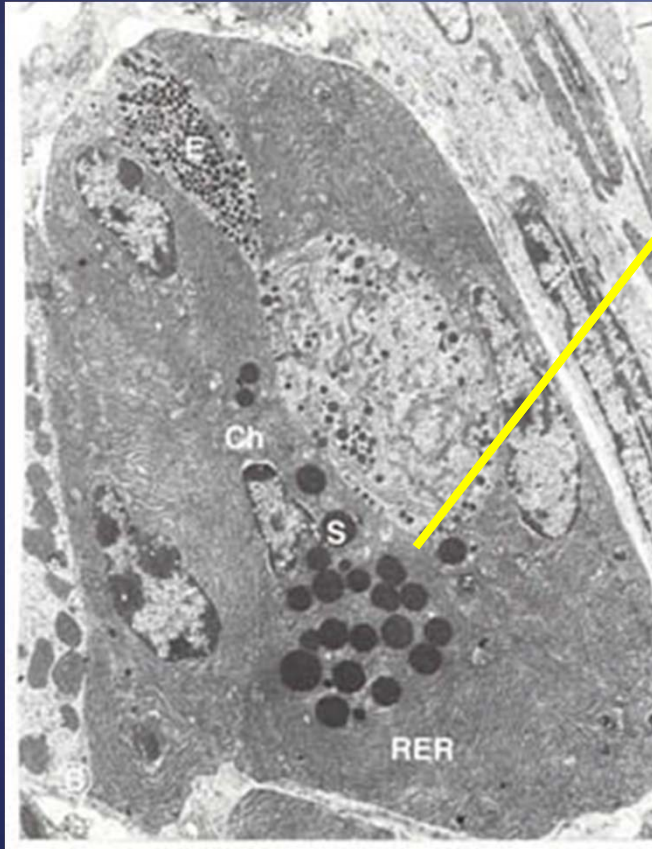


# Enterochromaffin (EC) cells





# Enterochromaffin (EC) cells



Serotonin (5-HT)

↑ blood flow

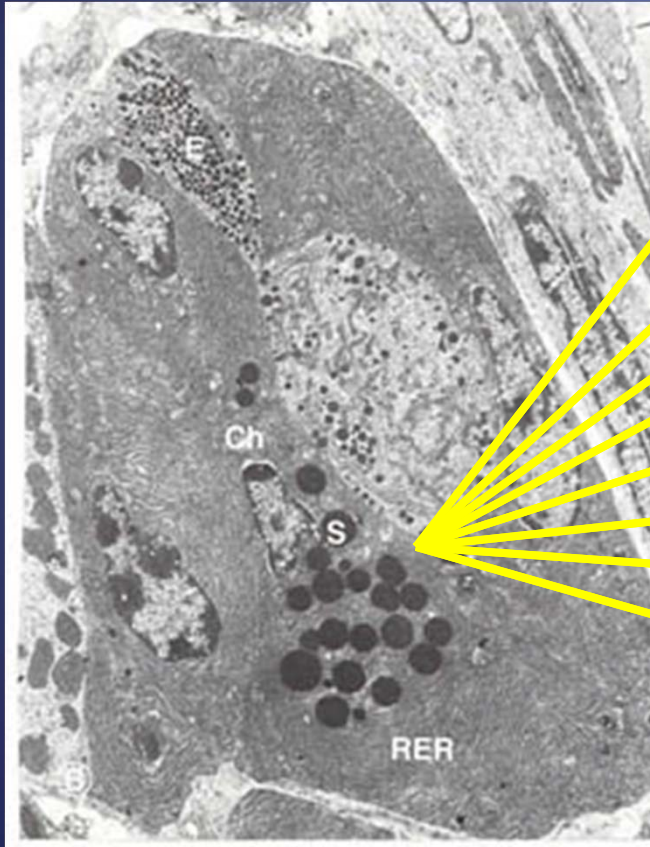
↑ secretions

↑ peristalsis

Metabolized &  
inactivated by liver

Measure in urine – 5HIAA

# Enterochromaffin (EC) cells



Serotonin (5-HT)

Melatonin

Substance P

Guanylin

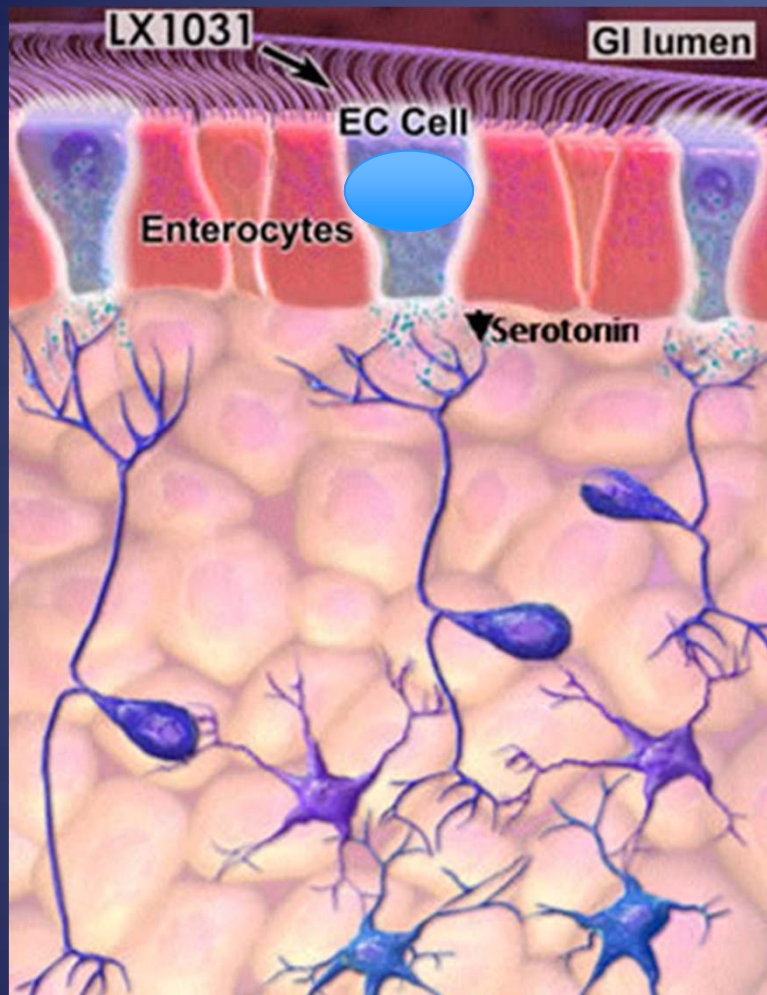
Histamine

Bradykinin

Chromogranin

Growth factors

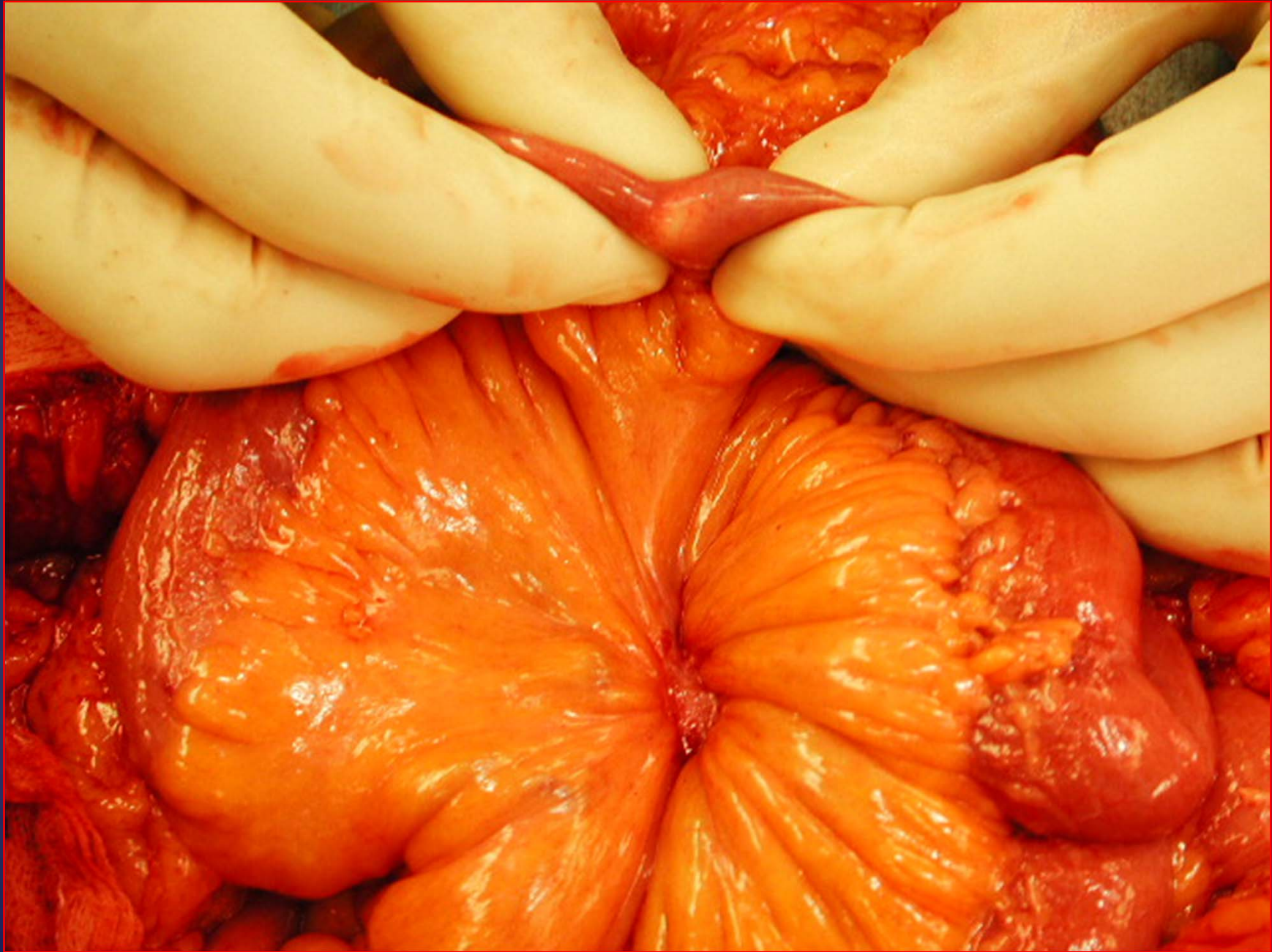




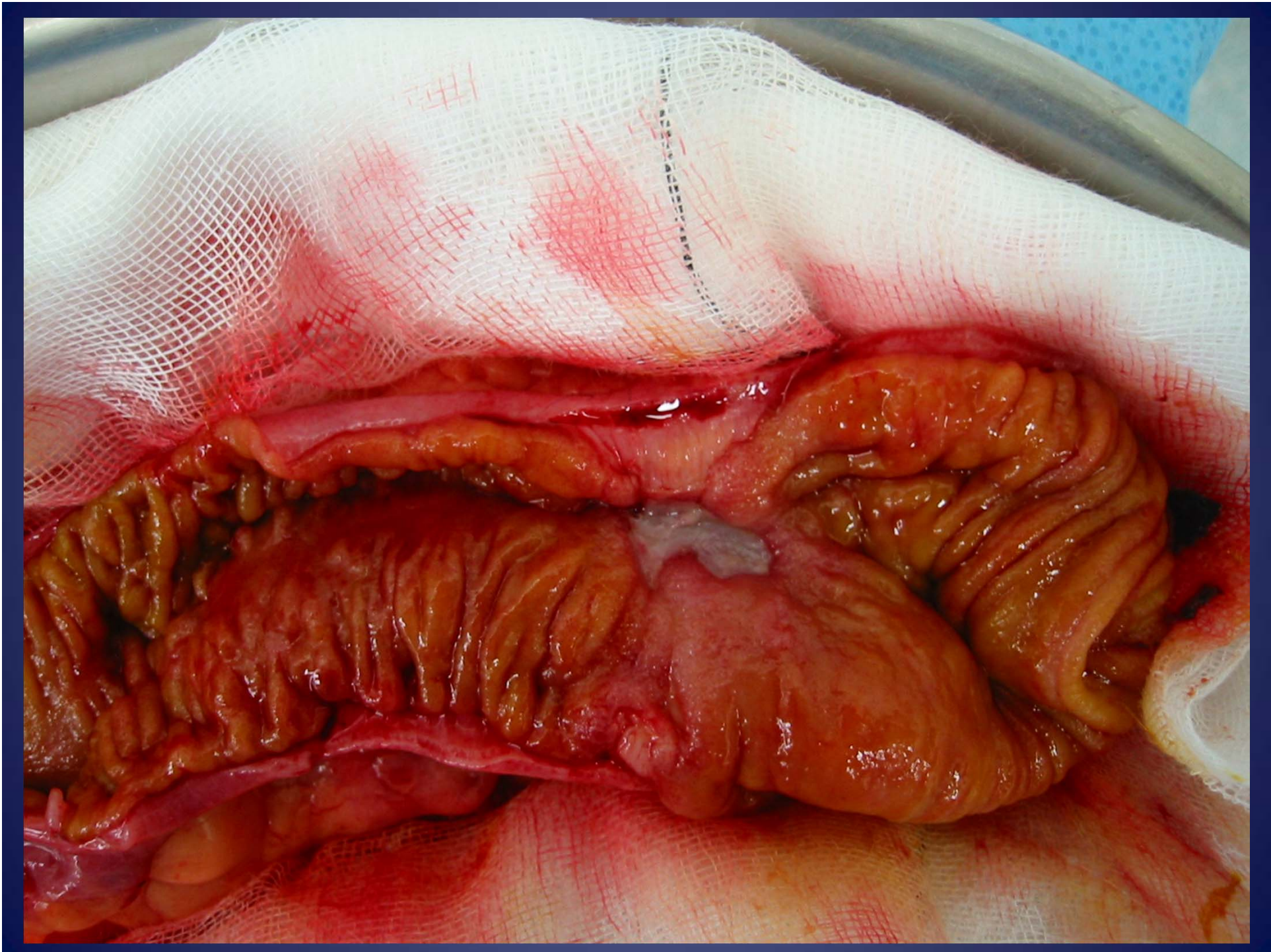
Intestinal contents  
Adjacent nerves

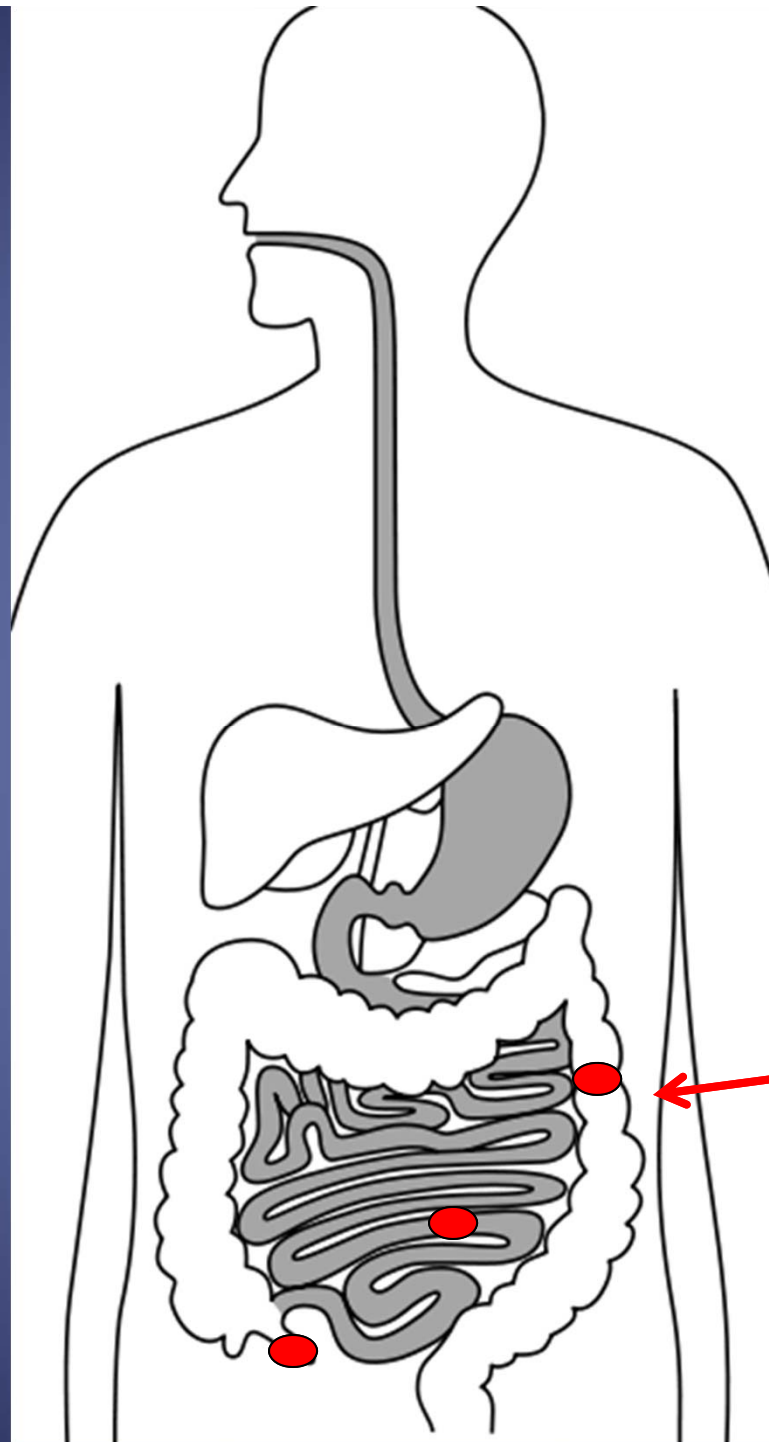


Somatostatin  
- (octreotide,  
lanreotide, LAR)



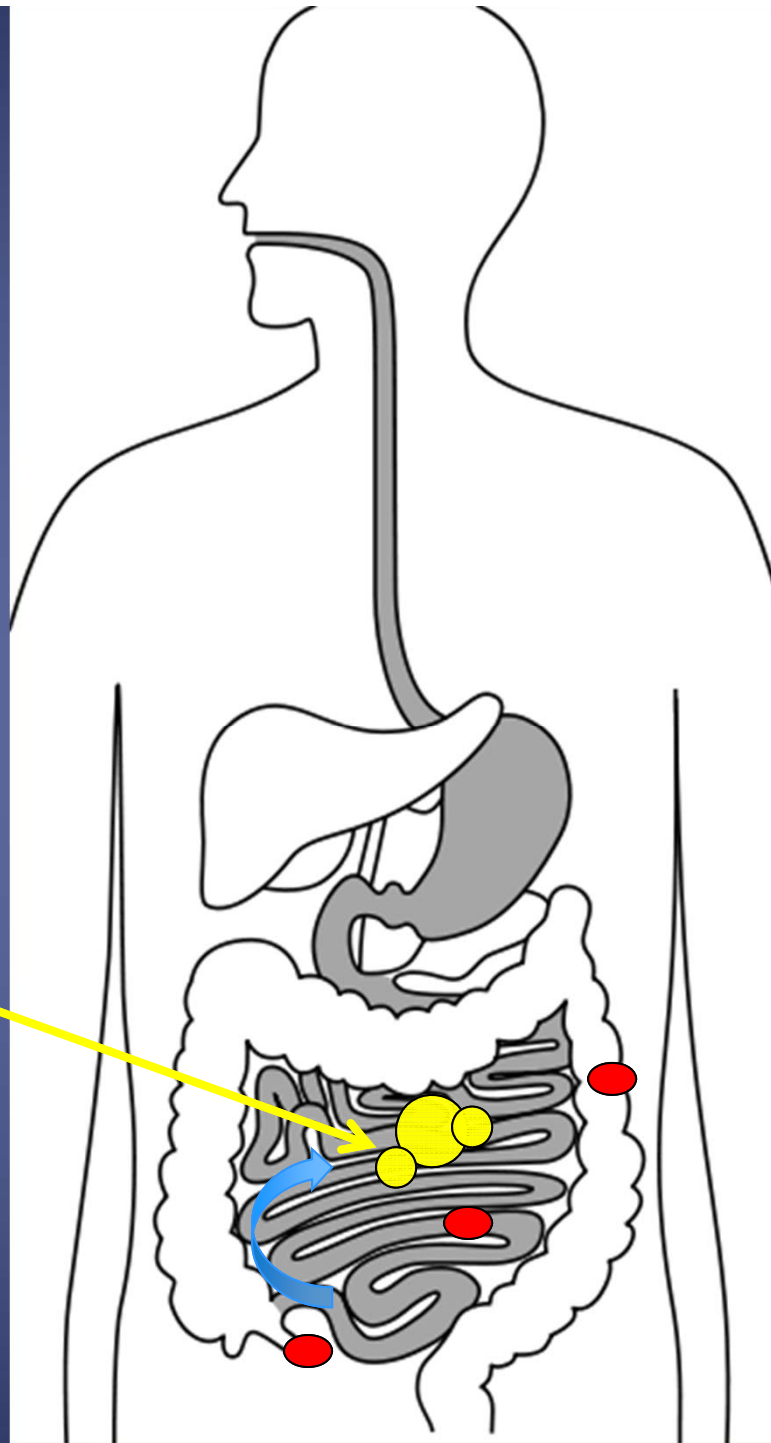






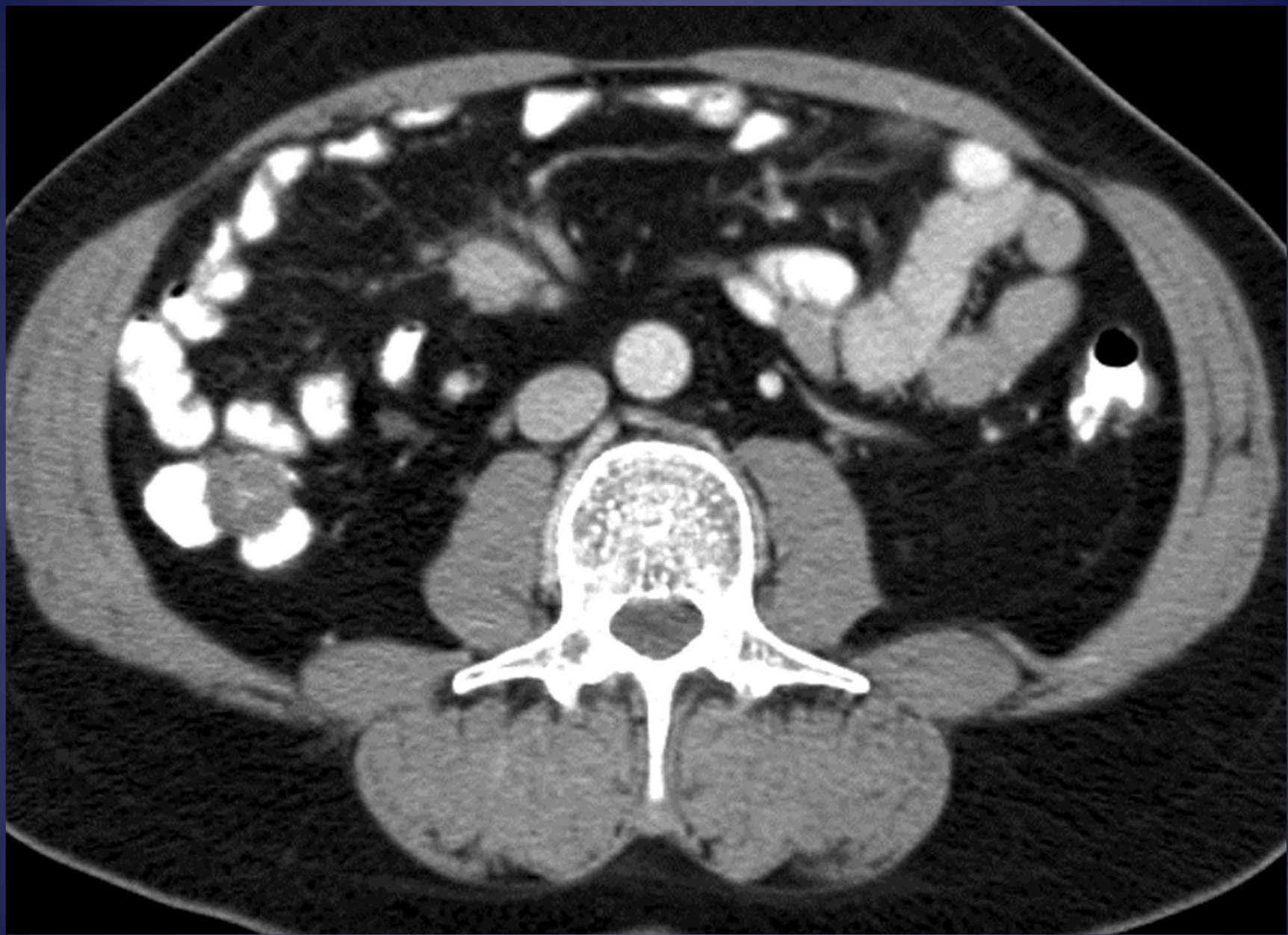
Primary  
Tumours

Mesenteric  
Lymph nodes



Primary  
Tumours

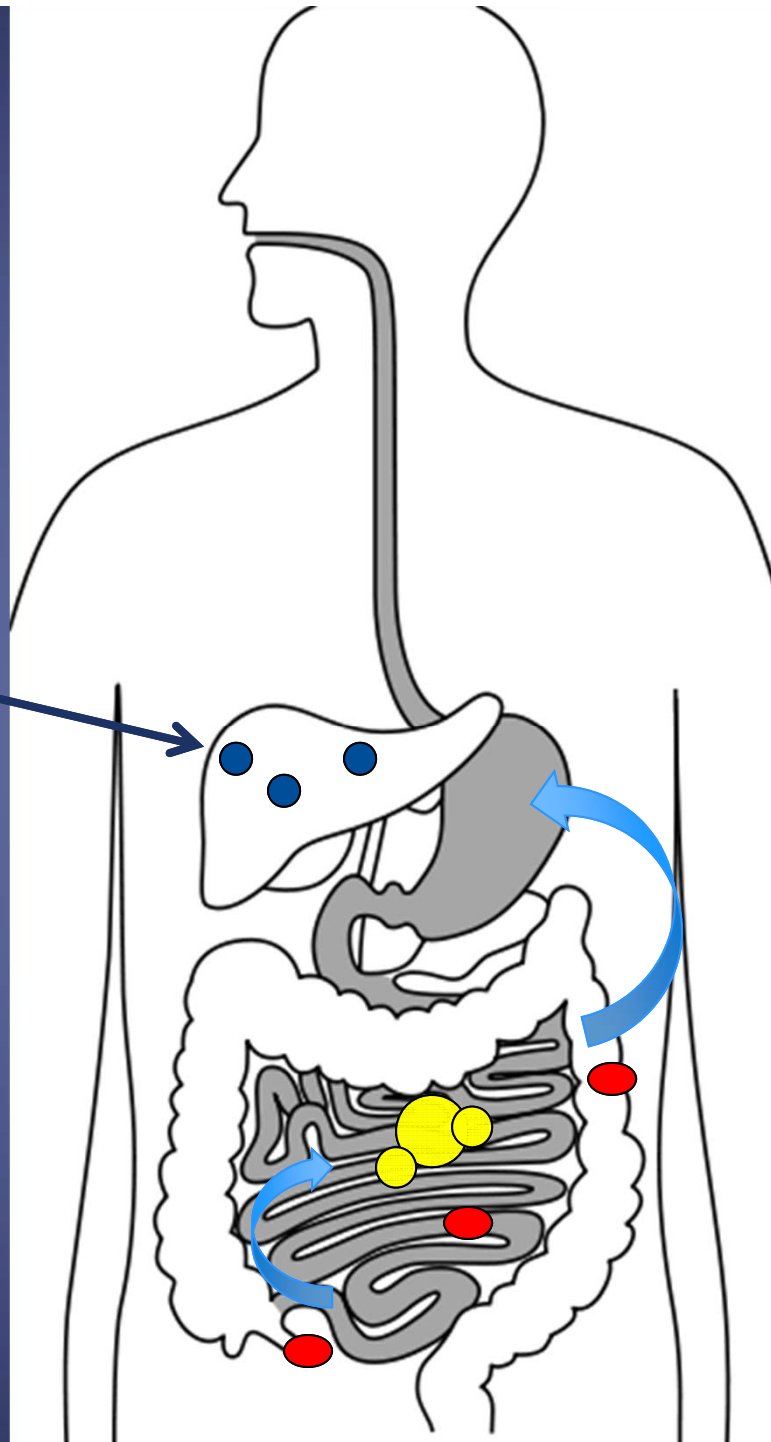




Liver  
Metastases

Mesenteric  
Lymph nodes

Primary  
Tumours







# Carcinoid Syndrome

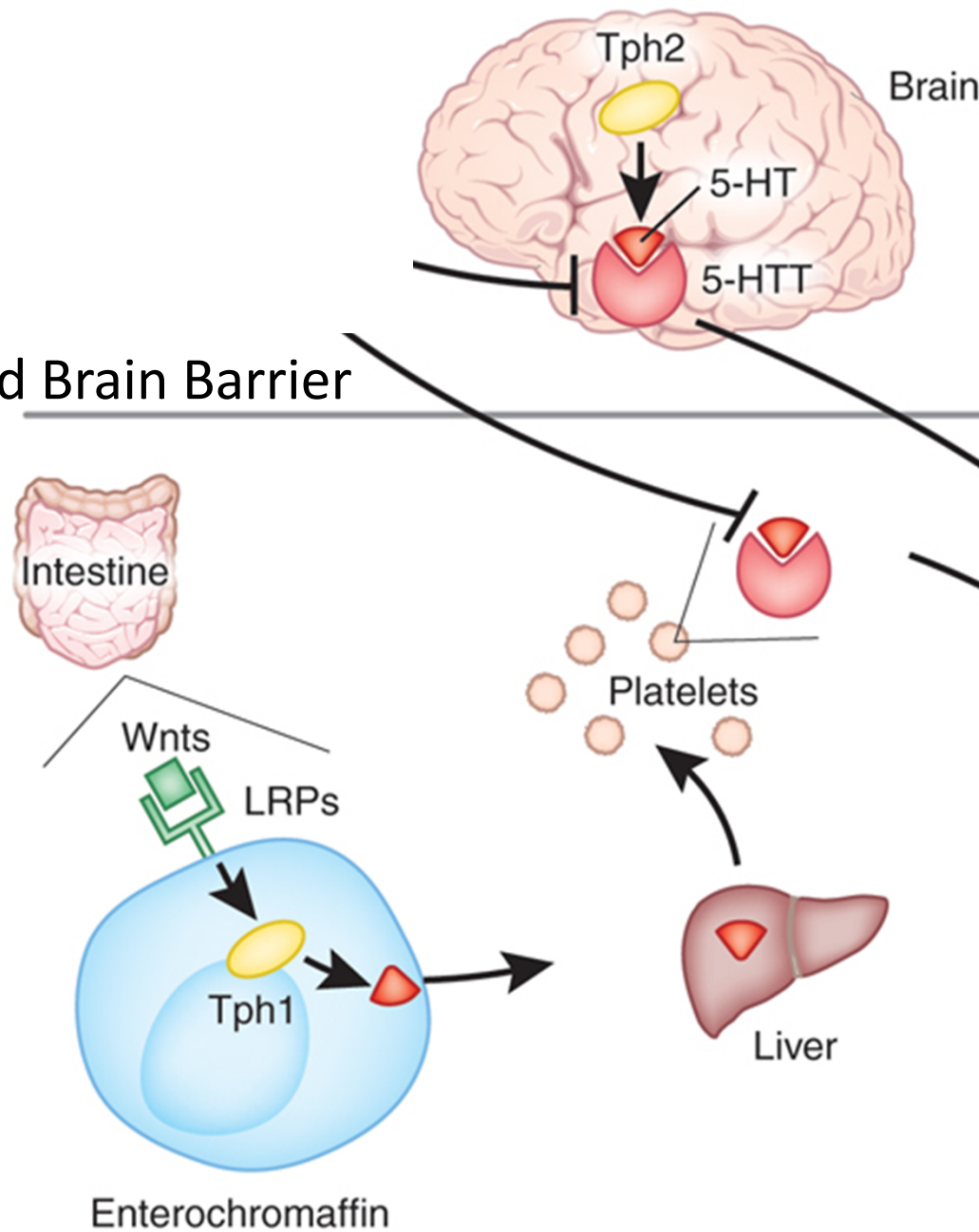
- Flushing of skin
- Diarrhoea
- Bronchospasm / wheezing
- Heart valve fibrosis
  - Tricuspid and pulmonary valves



# Carcinoid Syndrome

- Tests
  - 24 hr urine 5-HIAA
  - Serum Chromogranin-A
- Medical treatment
  - Octreotide / lanreotide / LAR

## Blood Brain Barrier





# Research study - Calgary

- Patients with Carcinoid Syndrome
  - ‘not themselves’
  - ‘difficulty remembering things’
  - ‘not as on the ball as they used to be’
  - ‘finding work difficult’
- ? symptoms of cognitive difficulties


# Research study - Calgary

- 21 patients – all small intestine NET with liver metastases & carcinoid syndrome
- Cognitive symptom questionnaire
- Formal neurocognitive testing by psychologist

# Symptom Assessment

- 38-Question standardized Multiple Ability Self-reporting Questionnaire (MASQ)

Cognitive Domain	Symptom score
Language	1 – 5
Attention / Concentration	1 – 5
Visual-Perceptual function	1 – 5
Visual memory	1 – 5
Verbal memory	1 – 5

Asymptomatic    1  5    Most severe symptoms



## MASQ Symptom scores

Cognitive Domain	Score $\pm$ s.d. (1 - 5)	Healthy norms
Verbal memory	2.75 $\pm$ 0.5	<2.1
Attention / Concentration	2.41 $\pm$ 0.7	<2.0
Language skills	2.31 $\pm$ 0.6	<1.8
Visual memory	2.30 $\pm$ 0.7	<1.8
Visual-Perceptual function	2.17 $\pm$ 0.6	<2.0

# Cognitive Testing

- Formal assessment - Neuropsychologist
- Battery of 6 standardized cognitive tests

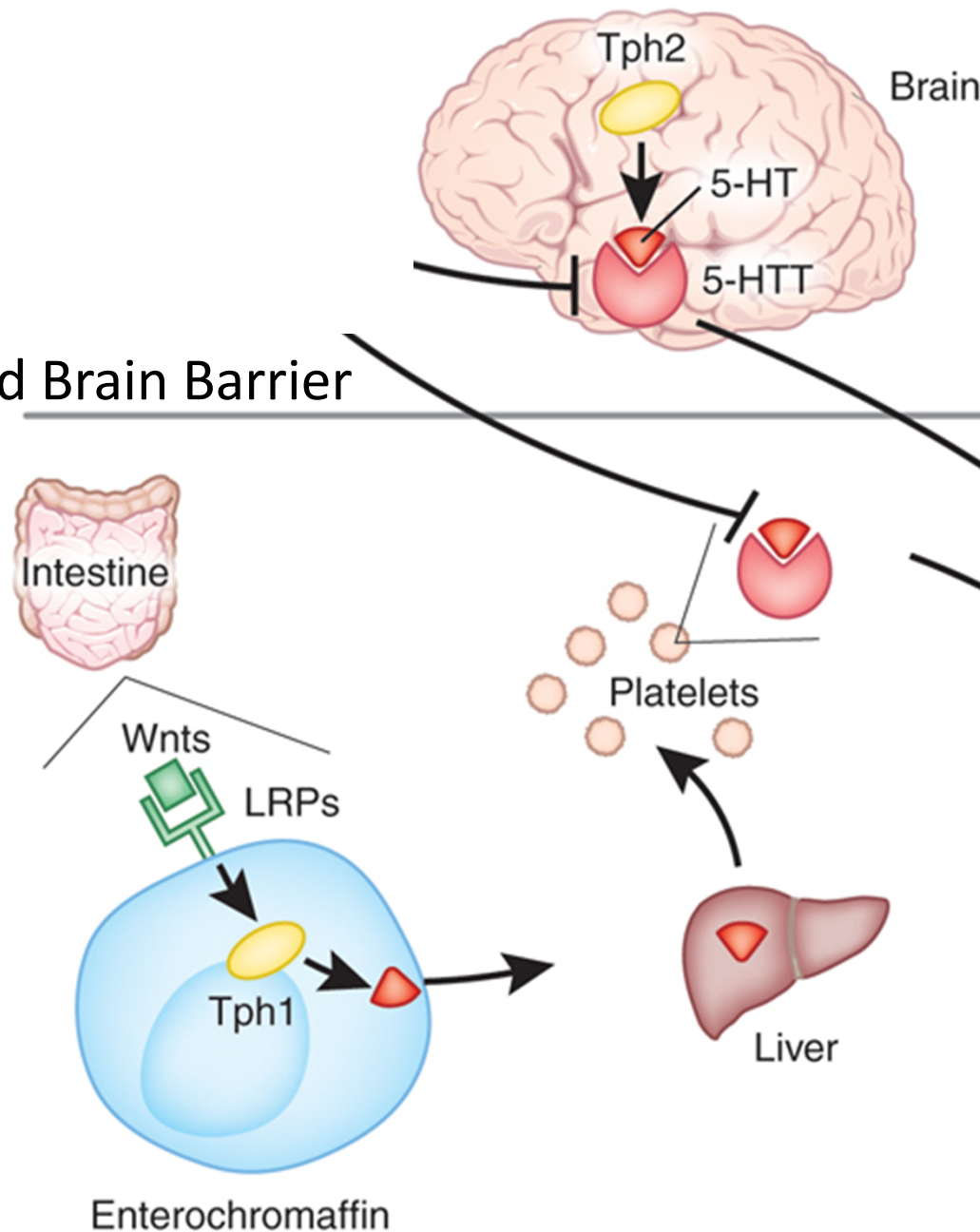
Cognitive Domain	Test
Attention / Concentration	-Trail making test -Verbal series attention
Execute skills / function	Wisconsin card sorting
Visual-Perceptual function	Complex figure recall
Language	Word & category fluency
Visual memory	Complex figure recall
Verbal memory	Selective reminding

## Summary of Results

Cognitive Domain	Symptom Scores	Cognitive testing
Attention/ Concentration	↑	Normal
Language skills	↑	Normal
Verbal memory - Immediate recall - Delayed recall	↑ ↑	Normal ↓
Visual-Percept Function	↑	↓
Visual memory	↑	↓

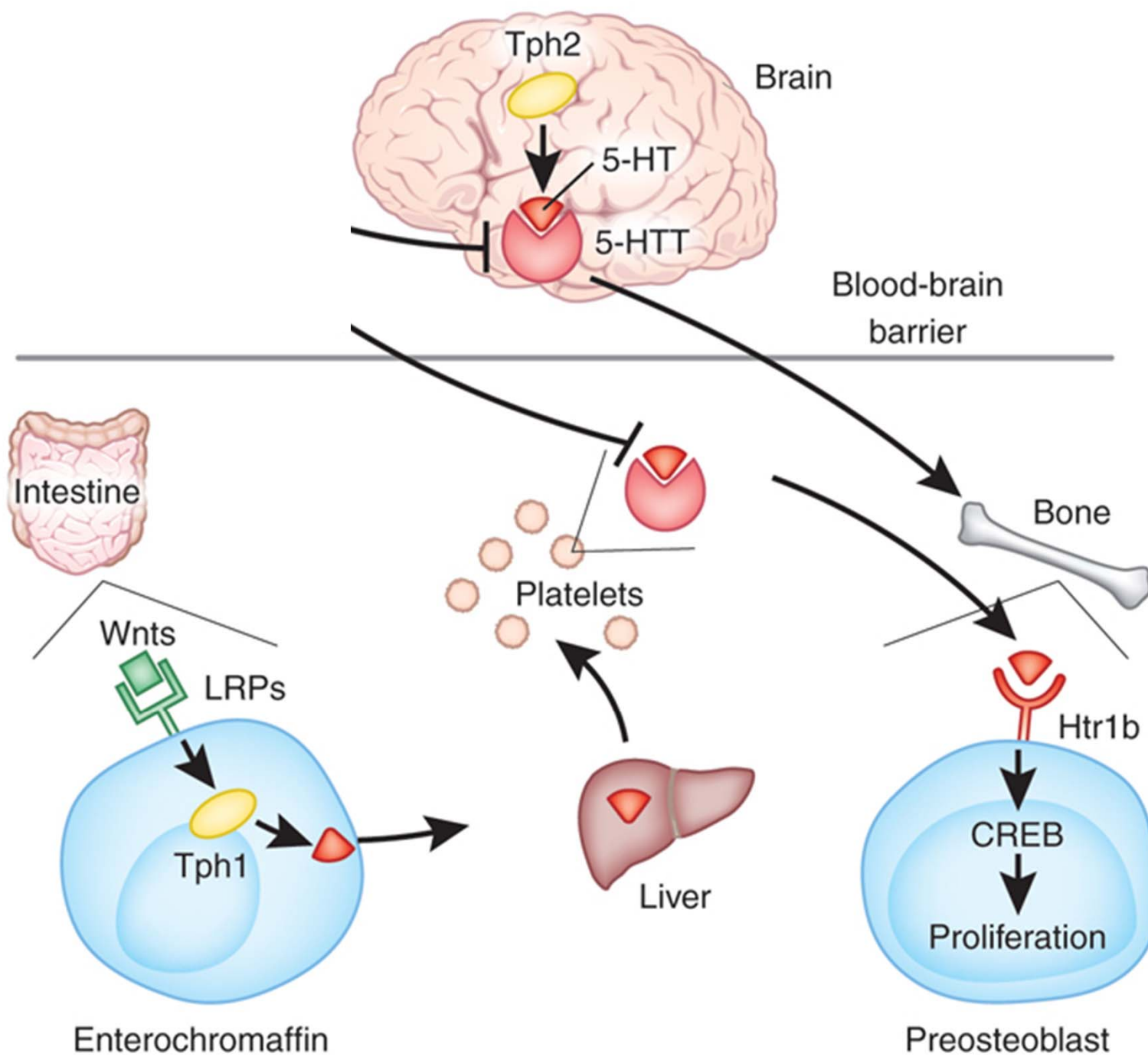


Blood Brain Barrier



## ? Cause

- Serotonin?
- Another tumour substance?
- treatment?
- malignancy?
- tryptophan depletion?



# Carcinoid Syndrome

- Our understanding of Carcinoid Syndrome is slowly improving
- Need for more research to more fully understand effects
- Plan for future research at St. Vincent's Hospital, Garvan Institute & University of Calgary



