Appendiceal tumors & pseudomyxoma peritonei

→ classification of appendiceal tumors

↓ epithelial

↓ Non epithelial.

Table 13.1 WHO classification of epithelial appendiceal tumours (Adapted from Ref. [6])

Epithelial tumours of the appendix	*Non-epithelial tumours*
Adenoma	Neuroma
• Tubular	Lipoma
• Villous	Leiomyoma
• Tubulovillous	Gastrointestinal stromal tumour
• Serrated	Leiomyosarcoma
Carcinoma	Kaposi's sarcoma
• Adenocarcinoma	Others
• Mucinous adenocarcinoma	Malignant lymphoma
• Signet ring cell carcinoma	*Secondary tumours*
• Small cell carcinoma	*Hyperplastic polyps*
• Undifferentiated carcinoma	
Tubular carcinoid	
Goblet cell carcinoid (mucinous carcinoid)	
Mixed adenoneuroendocrine carcinoma	
Others	

Epithelial tumors

↓

↓ Non infiltrative

(No invasion of muscularis propria)

↓ Low grade Appendiceal Mucinous neoplasm (LAMN)

↓ High grade Appendiceal mucinous neoplasm (HAMN)

(LAMN) — (HAMN) → >50%. extracellular mucin

↓ Infiltrative

↓ Adenocarcinoma

→ Mucinous (MACA) >50%. extracellular mucin

→ Signet ring cell carcinoma >50%. signet ring cells

→ Small cell

→ Undifferentiated

→ evaluation :

- usually incidental finding at appendicectomy.
- If found pre op → tumor markers — CEA
  CA 79.9
  ± CA 125.

- Non operative management of appendicitis done

↓

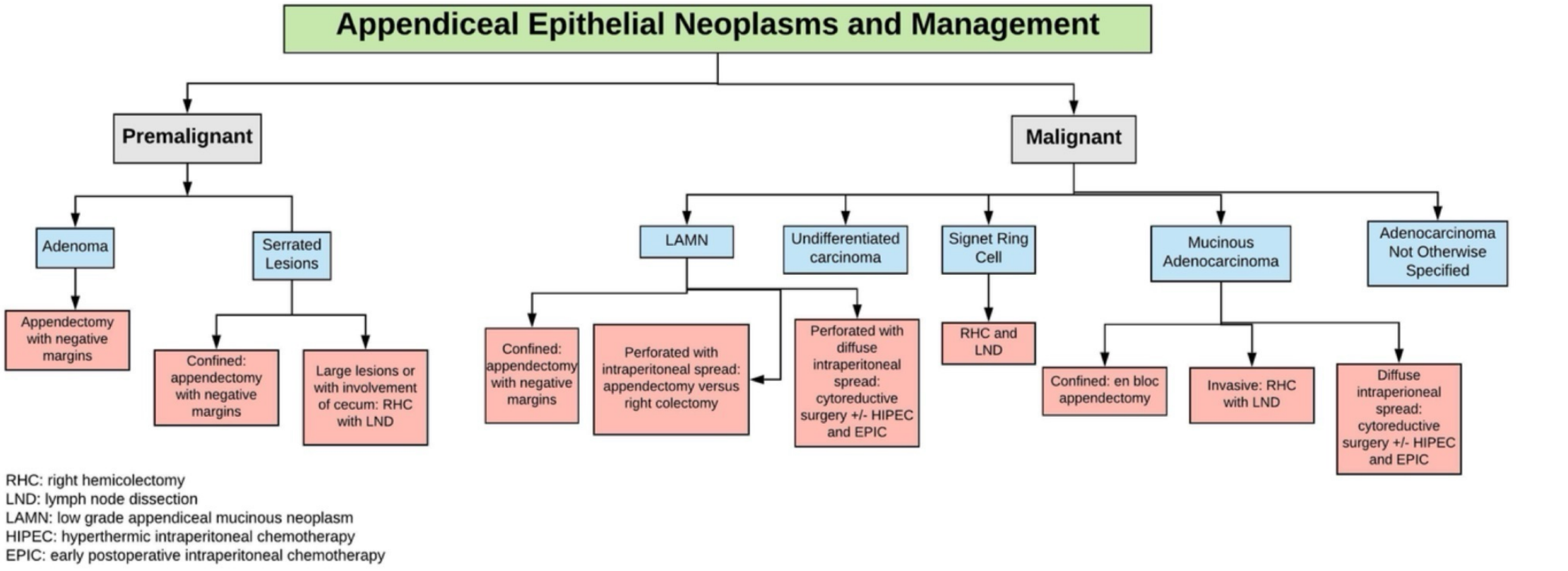
Repeat CT after 6 months to document Resolution of findings.

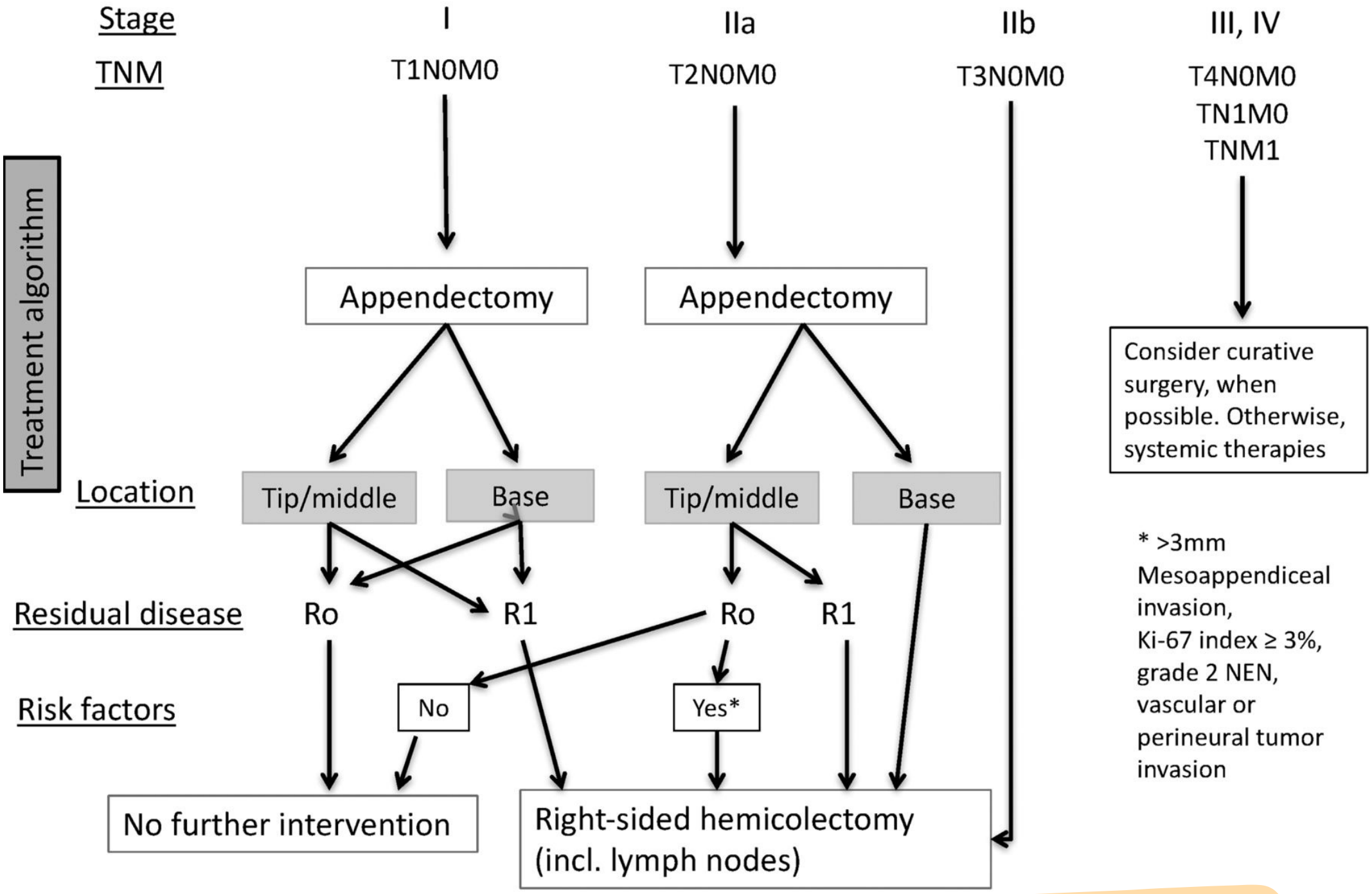
- Screening colonoscopy in all suspected patients of adeno CA appendix. to r/o Synchronous colonic malignancy.

↓

* Negative colonoscopy does not rule out appendiceal cancer.

→ management :





Treatment algorithm for NET of appendix

→ management of tumors found after appendicectomy:

Non invasive mucinous tumors

CAMN

↓

Active surveillance ± tumor markers, CT every 6-12 mo × 5-10 years.

Adenocarcinoma, But negative margin

↓

Second look laparotomy 6 months after primary Sx

No peritoneal disease

↓

Prophylactic cytoreduction

Peritoneal disease (+)

↓

Complete CRS + HIPEC. ± mitomycin C.

* Prophylatic Sx for Appendicular Adenocarcinoma:

- Greater & lesser omentectomy
+ Sampling of appendiceal nodes
+ Bil oophorectomy.

If positive,

(+) Right hemicolectomy.
ē conv + CVL

→ Pseudomyxoma Peritonei:

Presence of free or organised mucin with / without neoplastic cells in the peritoneal cavity following the typical pattern of redistribution.

90% → from appendiceal mucinous neoplasm.

6% → Mucinous carcinoma of ovary, GB, CRC, Pancreas, lung, breast.

* pathogenesis of PMP:

neoplastic transformation of Goblet cells

↓

Mucin production

↓

Tumor rupture & release of mucin

↓ high molecular weight heavily glycosylated proteins called MUC proteins

↓

Tumor cells lack cell adhesion molecules

↓

follow peritoneal redistribution phenomenon.

* Types of mucin
  - Membrane associated
  - Secreted
    - Gel forming → Colonic mucin MUC-2 is secreted in PMP.
    - Non Gel forming

→ Classification of PMP:

① Ronnet et al classification

Benign — DPAM — Disseminated Peritoneal Adenomucinosis.

Malignant
- PMCA — peritoneal mucinous carcinomatosis.
- PMCA-I — PMCA Intermediate group.

↓ Replaced by

② AJCC/WHO classification:

- Low grade PMP — <10% cellularity.
- High grade PMP — ↑ cellularity + atypical cells.

↓ Replaced by

(1) PSOGI Consensus classification:

Table 13.3 PSOGI expert consensus classification of PMP (Adapted from Ref. [7])

Lesion	Terminology
Mucin without epithelial cells	Acellular mucin
PMP with low-grade histologic features*	Low-grade mucinous carcinoma peritonei OR disseminated peritoneal adenomucinosis (DPAM)
PMP with high-grade histologic features*	High-grade mucinous carcinoma peritonei OR peritoneal mucinous carcinomatosis (PMCA)
PMP with signet ring cells	High-grade mucinous carcinoma peritonei with signet ring cells OR peritoneal mucinous carcinomatosis with signet ring cells (PMCA-S)

* Omental cake and ovarian involvement can be consistent with a diagnosis of either low-grade or high-grade disease

→ Diagnosis:

- H/o prior appendicectomy.
- Tumor markers – CEA, CA 19-9, CA-125.
- Features on imaging:
  - Scalloping of the surfaces of the liver and spleen by the mucinous deposits
  - Sparing of the small bowel serosa
  - Extensive omental involvement
  - Loculated intraperitoneal collections
  - Curvilinear calcifications
  - Fluid around the appendix or a mass in the appendiceal region [51]

* 18 FDG PET – may differentiate between grades based on uptake

- If imaging features are characteristic,

No role for Paracentesis & Diagnostic lap.

↓

If not, then D-lap to be done via midline access ports. (Avoid Laterally Placed ports) which can be excised.

→ Management:

- CRS + HIPEC: Multivisceral resection may be needed.
- CC-0 / CC-1 acceptable.

Table 13.4 Various drug regimens for HIPEC for appendiceal tumours

Regimen	IP drugs	IV drugs	Carrier solution	Duration
Mitomycin C based				
Sugarbaker regimen [88]	Mitomycin C 15 mg/m^2 Adriamycin 15 mg/m^2	5-Fluorouracil 400 mg/m^2 Leucovorin 25 mg/m^2	2 L of 1.5% dextrose peritoneal dialysis solution	90
Dutch high-dose mitomycin C regimen [89]	Mitomycin C 35 mg/m^2 17.5 mg/m^2 followed by 8.8 mg/m^2 at 30 and 60 min		3 L of 1.5% dextrose peritoneal dialysis solution	90

± EPIC + HIPEC ± Pemetrexed & cisplatin for peritoneal mesothelioma as it is a locally aggressive disease.