# General Data

- Age: 73y/o
- Sex: male
- Date of admission: 92-01-11

# Chief complaint

- progressive abdominal distension for 20 days

# Present Illness

- poor appetite,decreased intake, weight loss, short of breath at supine position, diffuse subtle abdominal pain, soft yellowish stool passage every few days.
- progressive abdominal distension ( ovoid shape ), and severe swelling of bilateral lower legs and feet.

# Past History

- Medical history：renal cyst found in MK87, keep f/u in our hospital until MK90
- Surgical history：peptic ulcer, s/p partial gastrectomy in 仁愛 hospital about 30 years ago
- subdural hemorrhage (?) after traffic accident s/p craniotomy in 博仁 hospital 7-8 years ago.
- cervical spondylosis at C5-C6 s/p EMG in TMCH, MK80

# Family **history**

- daughter: HBV carrier
- personal history
- Smoking:(+) smoking for 30 years 20 pieces/day, quited for 20 years
- Alcohol: (+) drinking for 30 years, quited for 20 years

# ER vital signs and PE

- afebrile ( 36.4C )
- Elevated blood pressure ( 161 / 73 mmHg ).
- shifting dullness, lower abdominal tenderness without accurate localization.

# Lab data

- anemia ( Hb 7.7 ), thrombocytosis ( PLT 638k ),
- no leukocytosis ( 6460 ).
- abdominal CT was arranged
- ( but internal bleeding, hepatoma rupture, and intra-abdominal abscess couldn't be ruled out ).

# Impression

- 1 intra abdominal tumor, nature ?
- suspect pseudomyxoma ( most likely )
- suspect intra-abdominal abscess

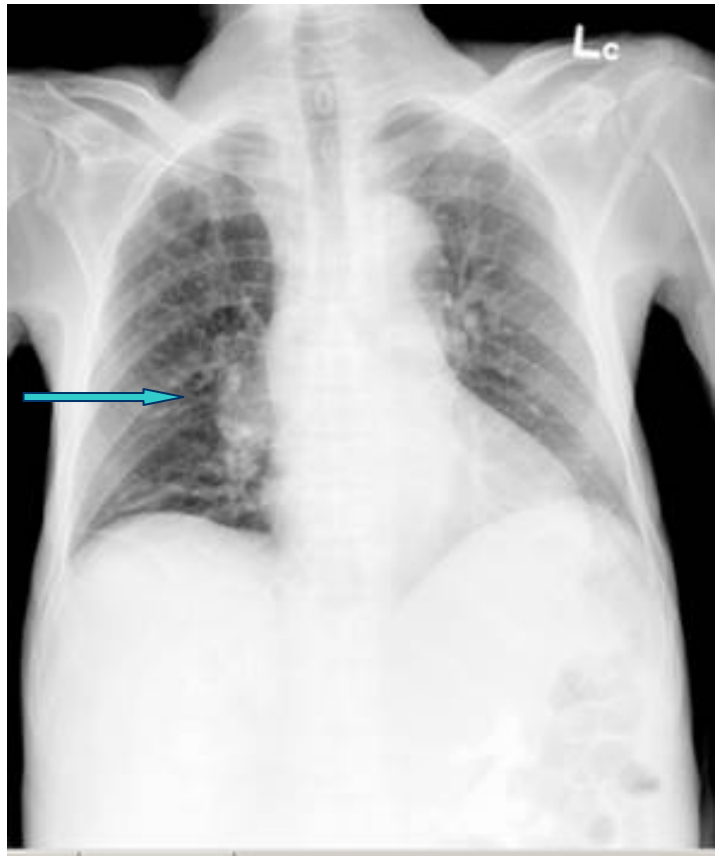
- 2 anemia, microcystic type, cause ?
- suspect iron deficiency anemia
- suspect infection-related
- suspect malnutrition-related

# Evaluation

- 1-11 Hb: 7.7, MCV: 78.0,
  WBC: 6460, Neut:81.5%, Plt: 638000
- GOT:19, GPT:19 BUN:17, Cr:0.8
- 1-12 Albumin:2.0,
- 1-13 CEA: 2.9 (0~4.6), AMY:15 Lip:8.0

# Evaluation

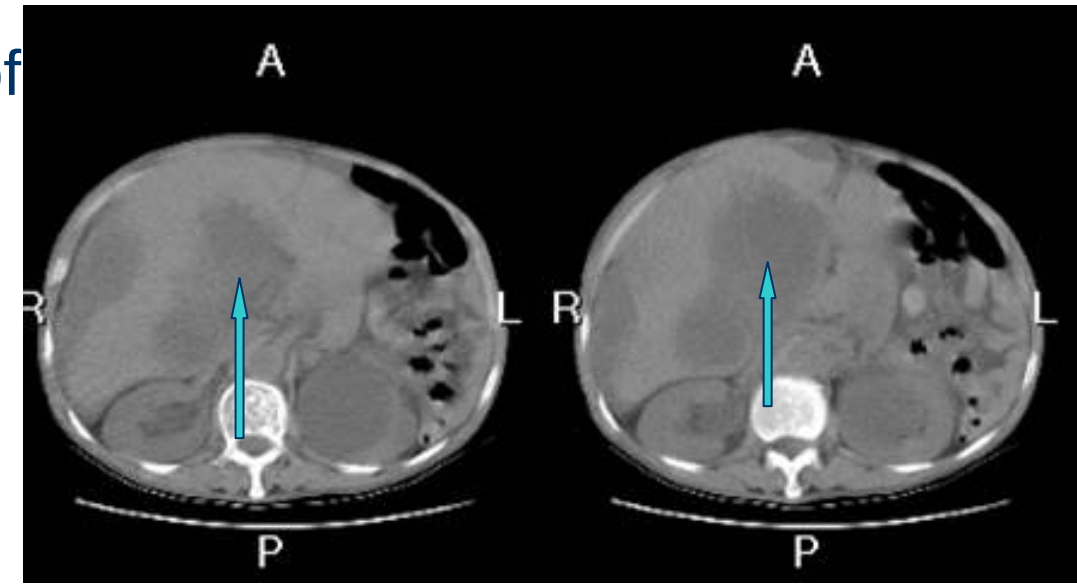
- 01-11CXR
- Right hilum nodule enlargement.



# Evaluation

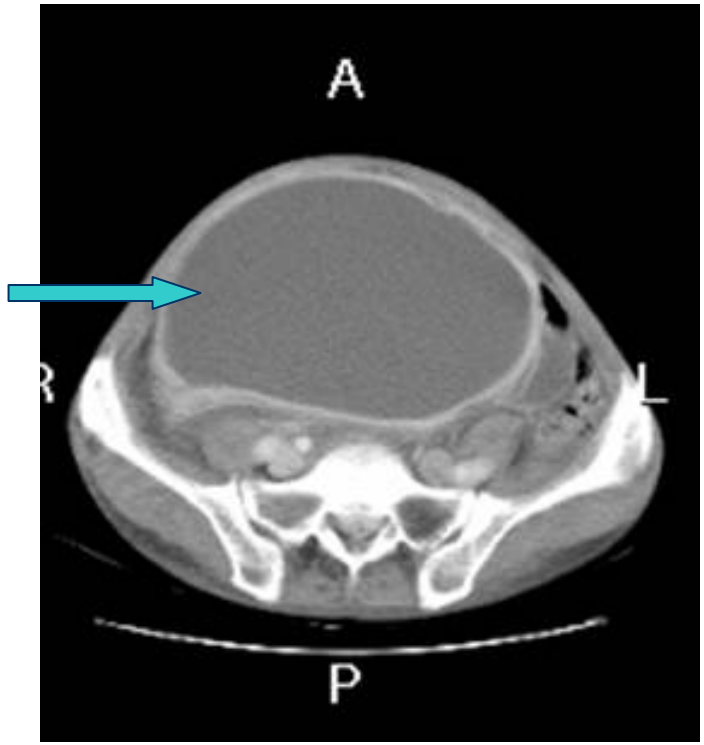
- Precontrast CT scan
- Irregular low density cystic mass located at right lateral side of liver with scalloping.

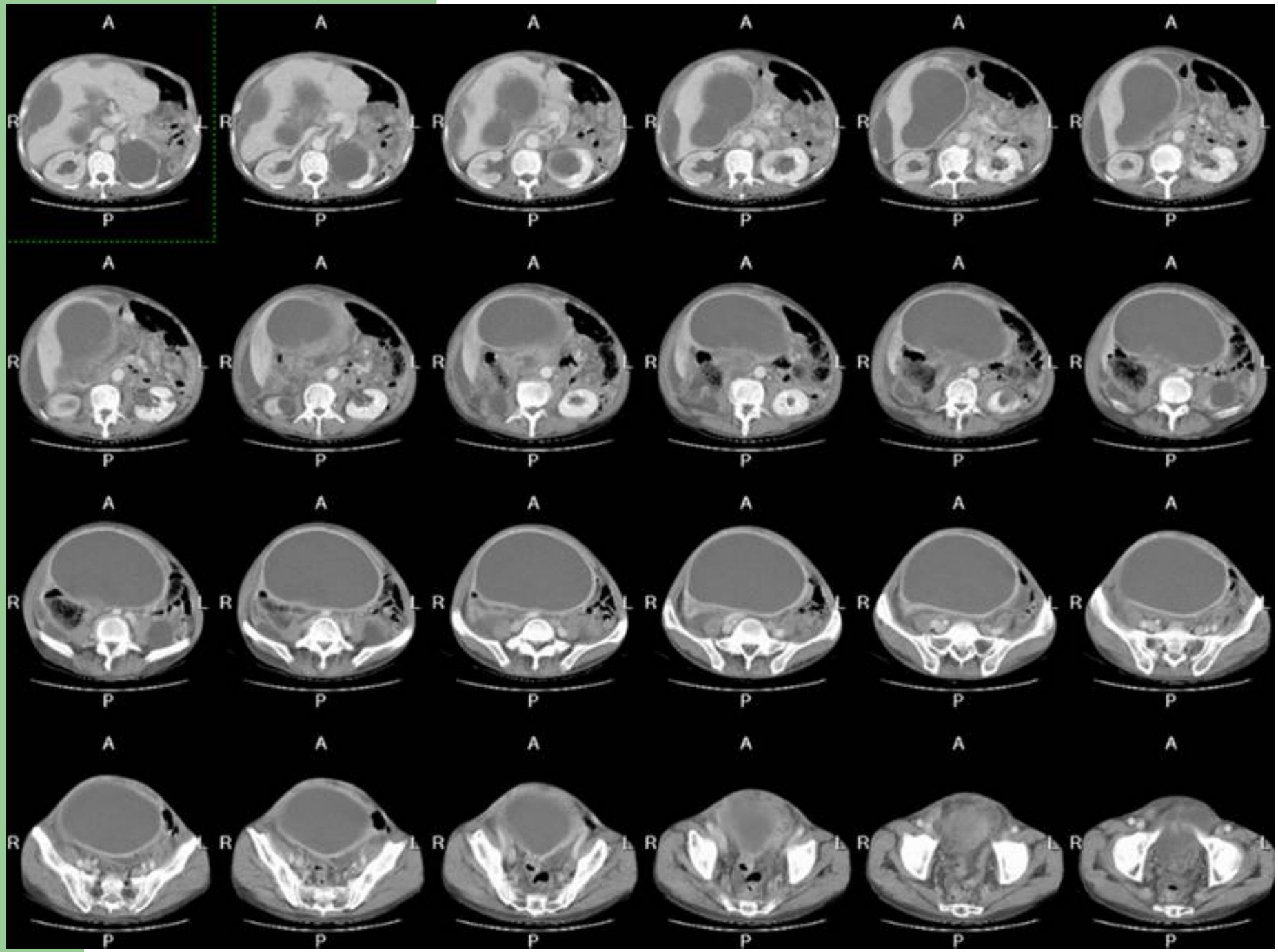
Left kidnet revealed a hydronephrosis or a cystic mass.



# Evaluation

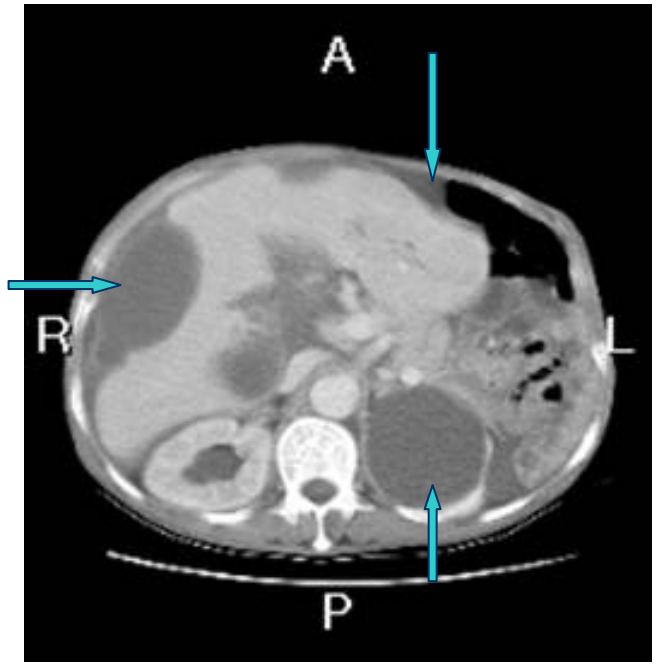
- Post contrast CT scan
- Low abdominal huge cystic mass with thick wall were noted.
- Measured about 20x12cm




A
R
L
P

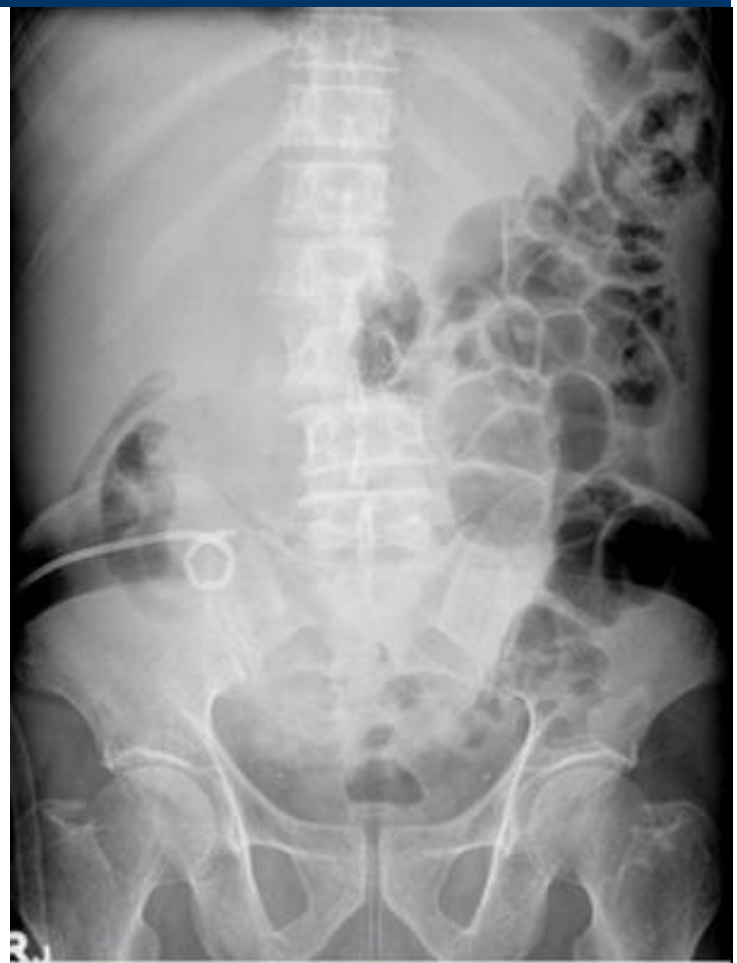
# Evaluation

- There were multiple unhanced subcapsule cystic mass noted.
- Huge left renal cyst were noted.
- At hilum, low density lesion were noted.



# Evaluation

- 01-14 KUB
- A central huge soft tissue mass was noted.
- Pig tail.



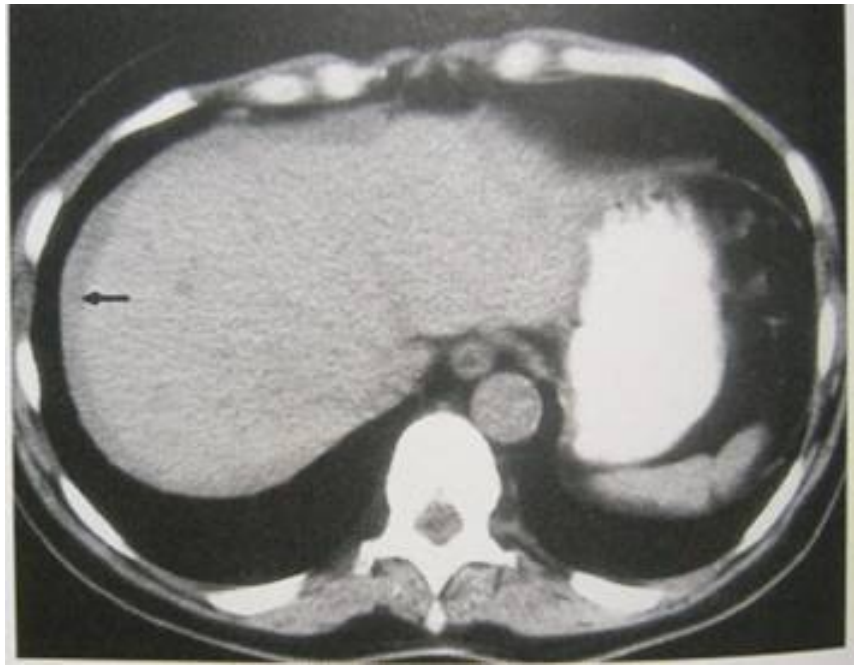
# Differential Diagnosis

# Differential Diagnosis

- Pseudomyxoma peritoni
- Abscess
- Mesothelioma
- Liver tumor
- Hematoma
- Ascites

# Mesothelioma

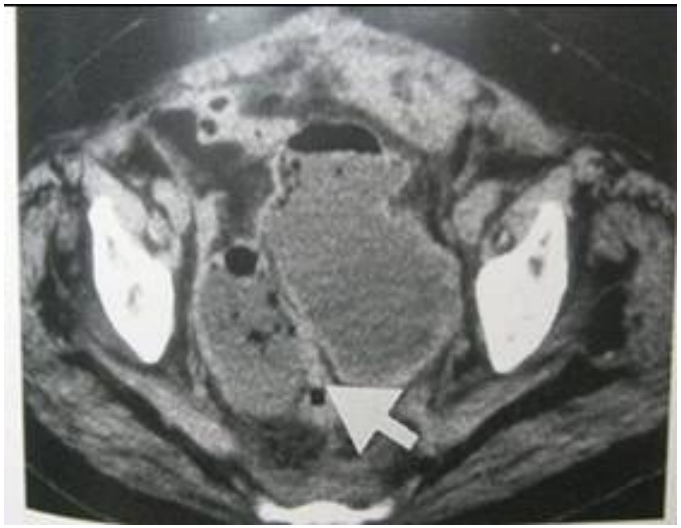
- Rare neoplasm arising in the peritoneal lining of the abdomen
- An area of soft tissue density within the abdomen
- May conform to the contour of the liver or produce a mass effect on adjacent structrure.



# Abscess

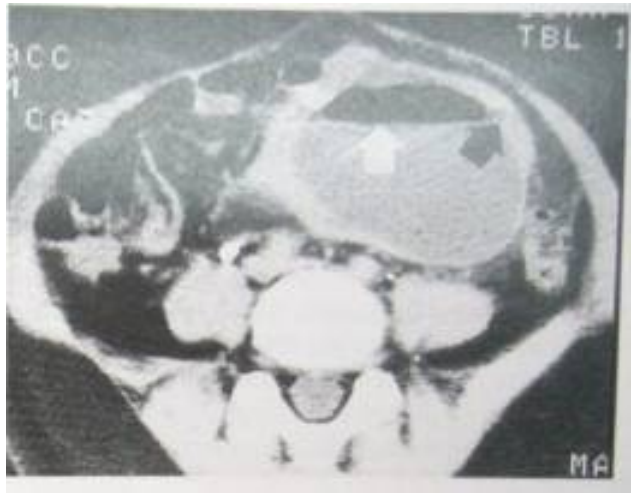
- 1/3 had gas—GI or bac.
- Well-defined or irregular margins.
- May contain multiple septations

# Abscess

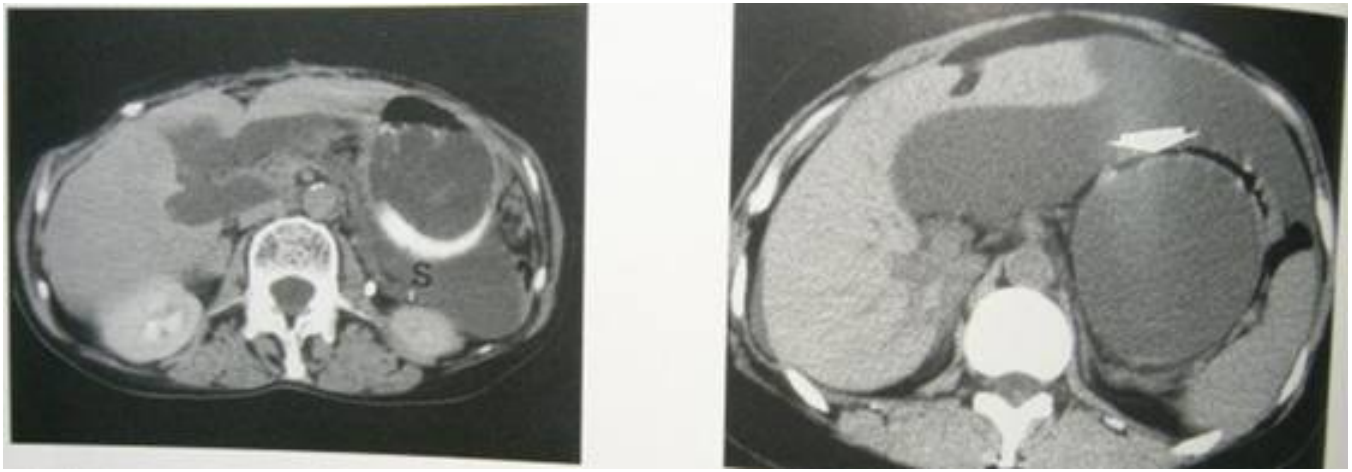


- Abscess filled with fluid and air, compressing the sigmoid colon.

- Air-fluid level



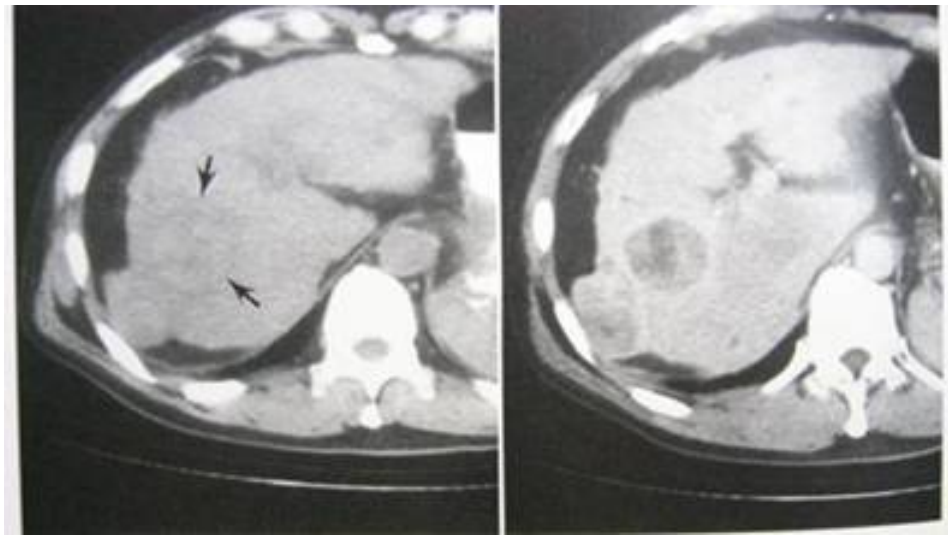
# Abscess



- L:fluid collection lesser sac splenic recess
- R:lesser sac, compressing the stomach anteriorly

# Liver tumor

- Multifocal HCC in cirrhotic liver
- L: unenhanced CT low-density lesions.
- R: contrast-enhanced CT
- Peripheral rim enhancement
- heterogeneous internal density

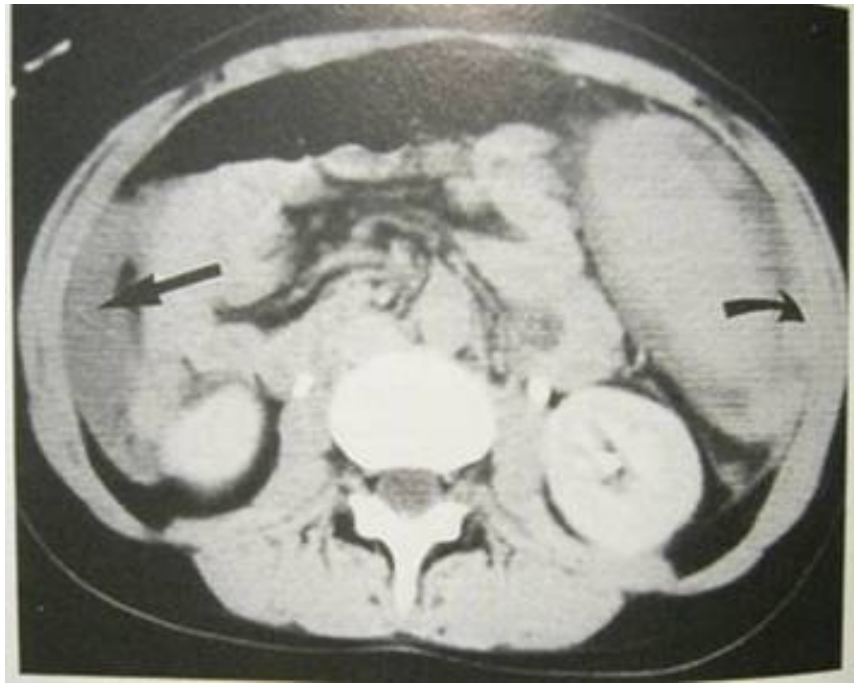


# Hematoma

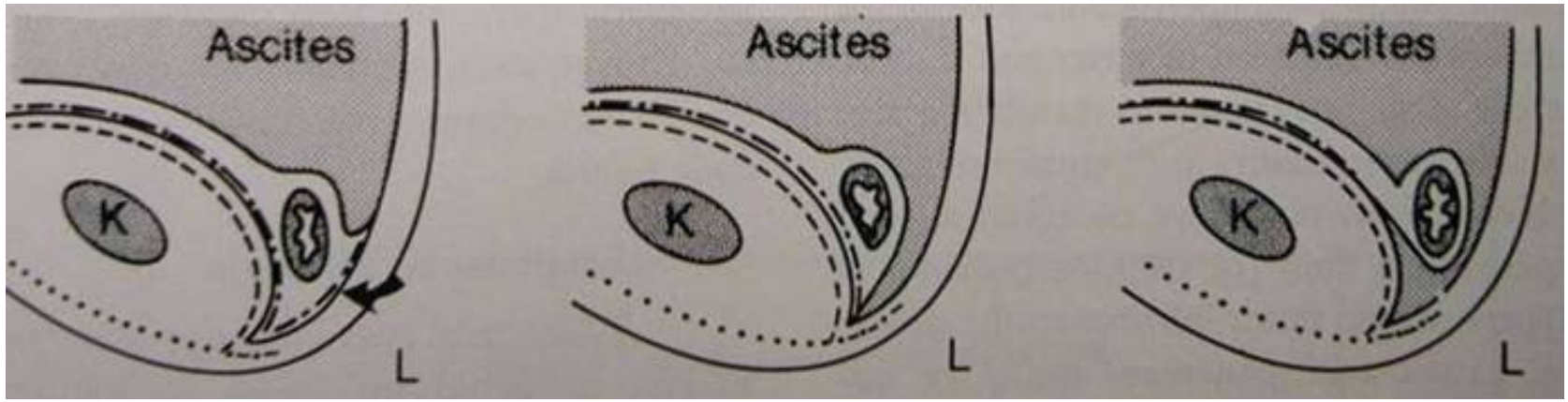
Right subhepatic space, pericolic spaces, pelvis, renal fascia

Fresh hemorrhage: high-density clot and serous fluid.

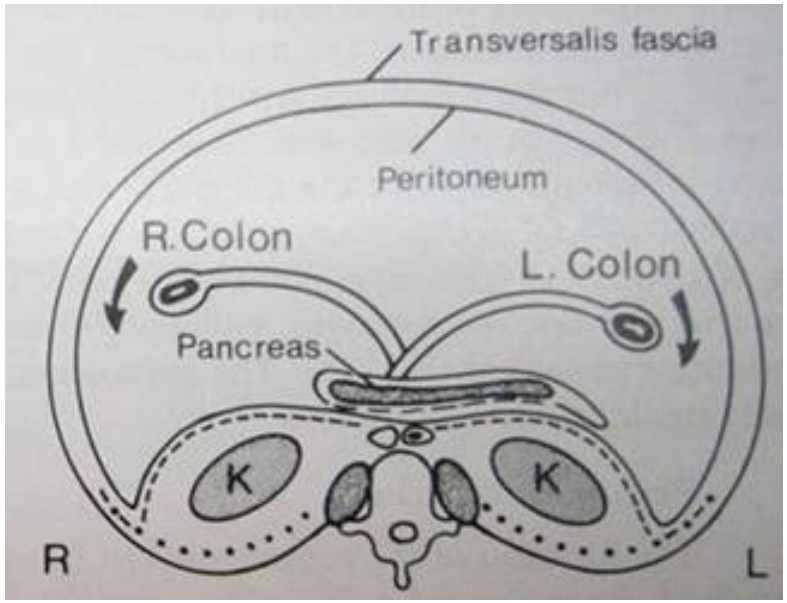
Lysed blood: low-density, similar to any other fluid

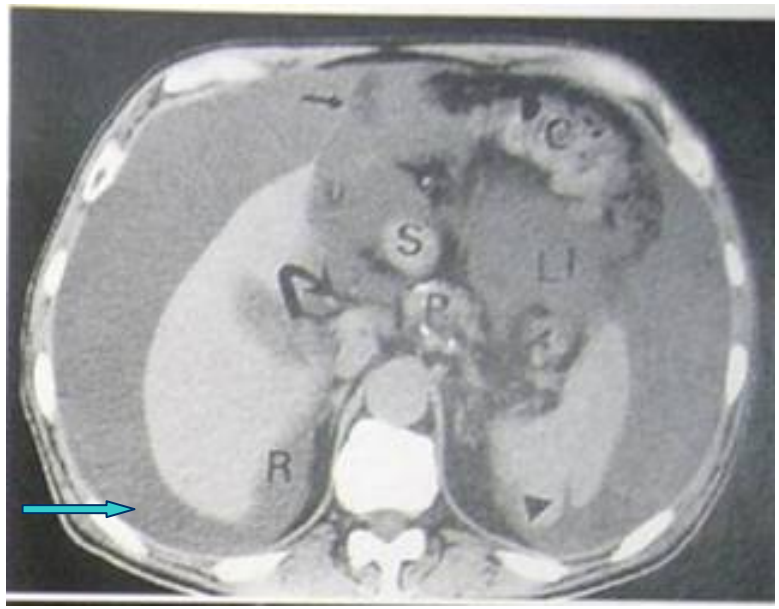


# Ascites



# Ascites

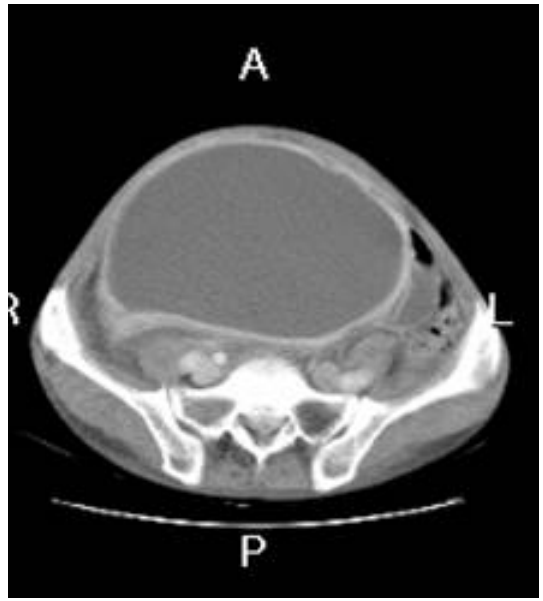

Transversalis fascia
Peritoneum
R.Colon
L. Colon
Pancreas
K
K
R
L


S
LI
R

# The abdominal mass

- Acute progressive abdominal mass→hematoma, fluid secreting tissue.

- In this case: vital signs stable, low density, capsule forming, homogeous, no gas.



# Operation:92-01-15

- 1.Intra-abdominal mass with mucoid content and fibrotic capsule.
- 2.Abscess cavity with mucoid content aboud 02*20cm.
- 3.Frozen section:inflammatory change
- 4.Debridment of multioculaters abscess inside abdominalcavity, right parabloic region, bubhepatic and subplueric.

# Pathology 92-01-19

- cyst wall:picture of fibrosis and granulation tissue in the thick cyst wall. Mucinous pools are noted in the cyst wall. There is fibrinous exudate coating on the inner surface. No lining epithelium is seen in this specimen.
- **INNER CONTENT (MUCUS) :shows fibrinoid necrotic debris. Frozen diagnosis: necrotic debris**
- **INTRAABDOMEN ABSCESS :show a picture of pseudomyxoma peritonei with a few well-differentiated mucinous columnar cells of intestinal type floating at mucinous material. Most peritoneal tissue reveals acute and chronic inflammatory cell infiltration, fibrotic cystic change and granulation tissue formation.**

# Operation finding:92-02-03

- 1.ascending colon tumor with serosal exposure and peritoneal dissemination (mucous content) about 10cm from ileocecal valve.
- 2.tumor size 8*8*8cm
- 3.shallow penetration to serosal layer.

# Pathology

- An ulcerative cauliflower-like colon tumor,
- measuring 8 x 7 x 4 cm in size, is located at 2.5 cm distal to the ileocecal valve.

# Discussion

# Pseudomyxoma peritoni

- **Diffuse dissemination of mucinous material derived from neoplasms.**
- 佔所有剖腹手術的千分之一以下。
- 50~70 y/o
- Male-- 闌尾的黏液囊腫，
- Female-- 卵巢的黏液樣腺癌。
- 10~20%: from GI tract Ex: stomach, colon, pancreas.
- Slowly progressive malignancy.
- Distant metastasis: rare.

# Symptoms and Signs

- **the most common symptom : gradually increasing abdominal girth.**
- **2nd most common symptom**
- **In males: inguinal hernia.**
- **In women: ovarian mass palpated at the time of a routine pelvic examination.**

- **Some of the earliest diagnoses are in patients in whom the disease was detected at laparoscopy performed for infertility. A laparoscopic diagnosis of mucinous appendiceal tumor is becoming increasing frequent.**
- **"jelly belly" :caused by copious mucus tumor production**

# Implant site

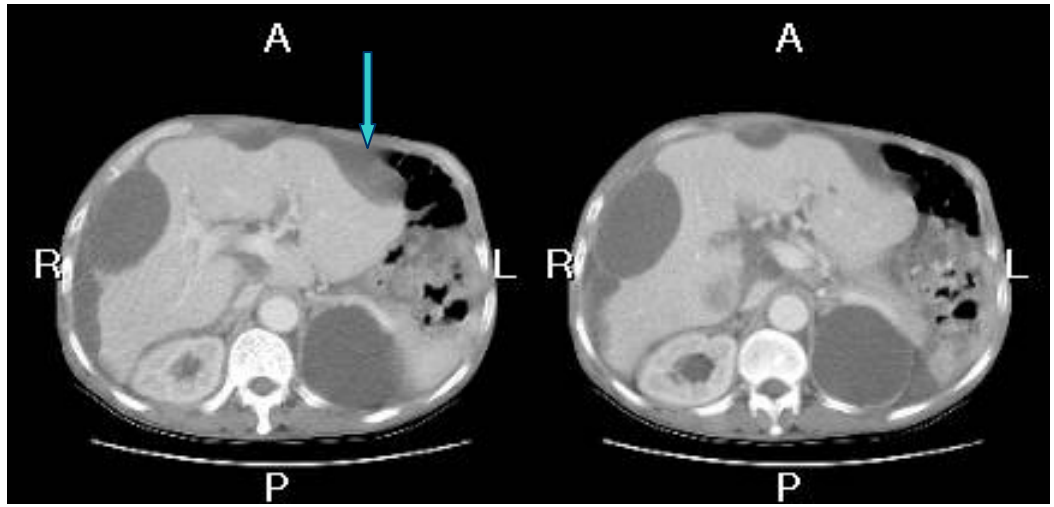
- Unlike carcinoma cells, peritoneal adenomucinosis cells accumulate at specific abdominal and pelvic sites.
- The factor that determines the localization : the absorption of peritoneal fluid.
- Bulky deposits : the greater omentum, lesser omentum and beneath the right hemidiaphragm.
- Gravity, Dependent portions of the abdomen and pelvis.

# the primary tumor

- **with pseudomyxoma peritonei, the primary tumor is usually inconspicuous and rarely causes symptoms because of its size**.

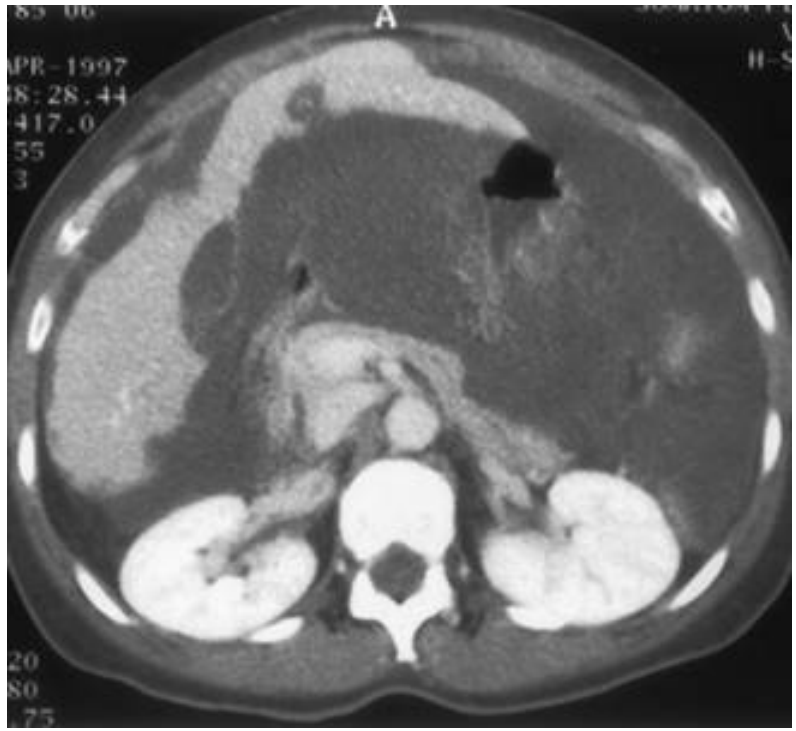
# Imaging

- 超音波檢查: 具回音性的腹水
- 腸氣並非自由地漂浮於腹水之中，反而被推向腹部後方，這點有別於其他病因引起的腹水。
- 電腦斷層檢查亦具診斷價值，最主要的發現是瀰漫性的低緻密度的腹腔內腫塊及腹水，有些病例並可看到這些腹水在肝脾的表面壓出海扇型的凹陷(scalloping sine)。



# Imaging

- due to mucinous cystadenocarcinoma of the ovary.
- lesser sac and greater sac
- scalloping of the liver margin.



# Pathology

- "disseminated peritoneal adenomucinosis" or simply "adenomucinosis".
- Cases of peritoneal carcinomatosis, regardless of the presence of abundant extracellular mucin, are excluded from this definition..

# Prognosis

- It is a deadly process.
- Although it does not metastasize via the lymphatics or blood stream
- The space required within the abdomen and pelvis for nutritional function eventually becomes replaced by mucinous tumor.
- Pseudomyxoma peritonei always results in the death of the patient unless definitively treated.

# **Treatment** :Cytoreductive Surgery

- **Because it is minimally invasive and yet extensively coats parietal surfaces, a series of peritonectomy procedures were developed.**
- **Stripping the parietal peritoneum and resecting structures at fixed sites that contain adenomucinosis.**

# Op finding

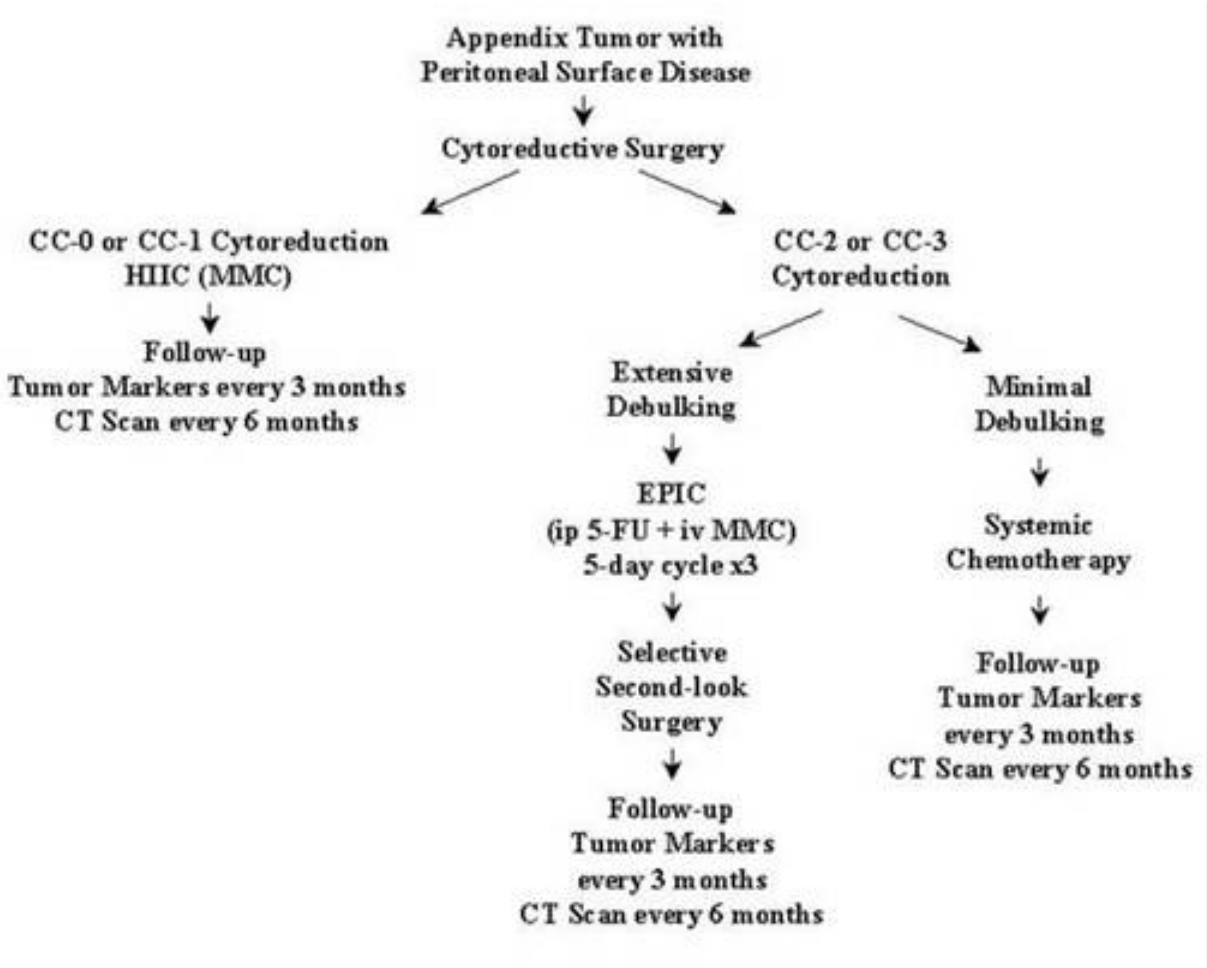
- Perhaps the most consistent observation during surgical exploration : complete or nearly complete absence of mucinous tumor on the intestinal surfaces.
- exceptions :the antrum of the stomach and pylorus, the ileocecal valve region, and the rectosigmoid colon within the pelvis.
- Reason:1. fixed to the retroperitoneum 2. not free to move → prevents mucinous tumor implantation on these surfaces.

# Treatment

- Direct administration of selected drugs into the peritoneal cavity.
- This pharmacologic advantage : peritoneum-plasma barrier.
- major problem :nonuniform drug distribution.

  → The use of Heated Intraoperative Intraperitoneal Chemotherapy (HIIC) after complete dissection of an adhesive process and before anastomoses are completed.

HIIC improves the drug penetration into tissue


Appendix Tumor with
Peritoneal Surface Disease
Cytoreductive Surgery
CC-0 or CC-1 Cytoreduction
HIIC (MMC)
Follow-up
Tumor Markers every 3 months
CT Scan every 6 months
CC-2 or CC-3
Cytoreduction
Extensive
Debulking
EPIC
(ip 5-FU + iv MMC)
5-day cycle x3
Selective
Second-look
Surgery
Follow-up
Tumor Markers
every 3 months
CT Scan every 6 months
Minimal
Debulking
Systemic
Chemotherapy
Follow-up
Tumor Markers
every 3 months
CT Scan every 6 months

***Thank you!!***