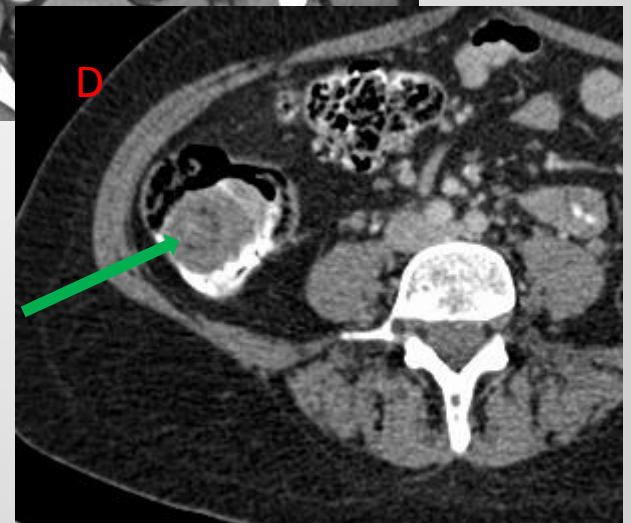
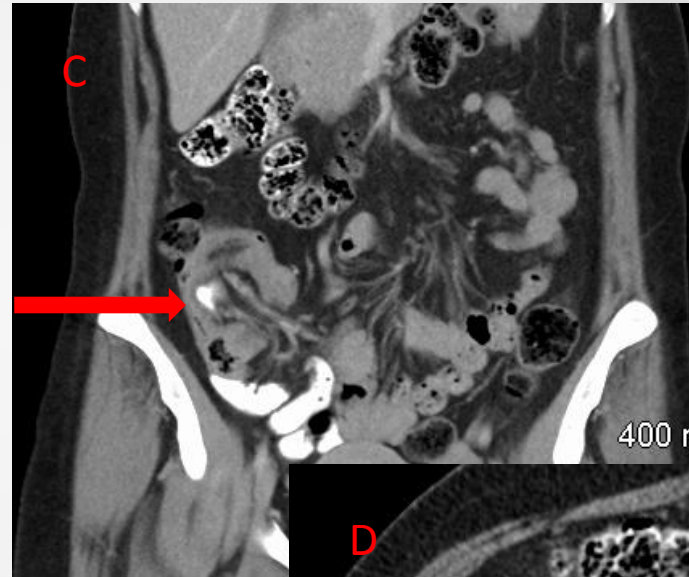
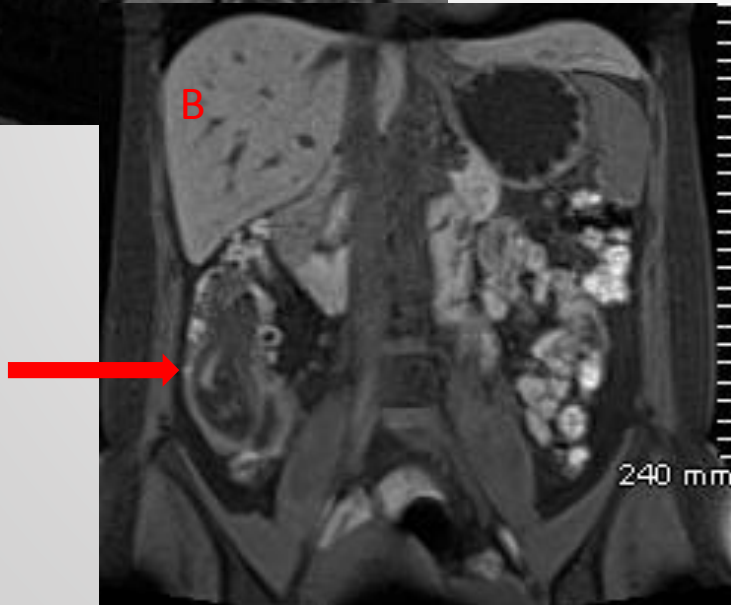
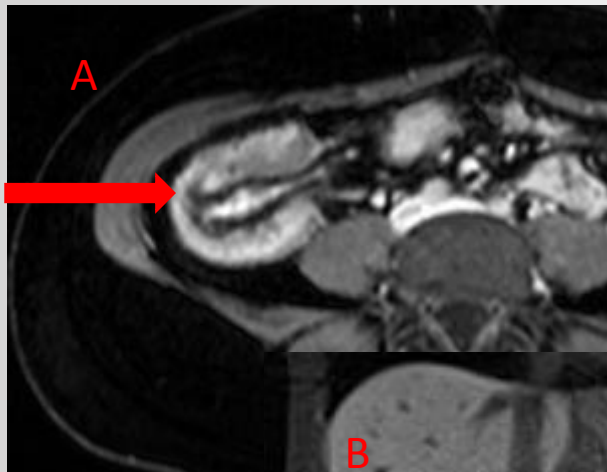
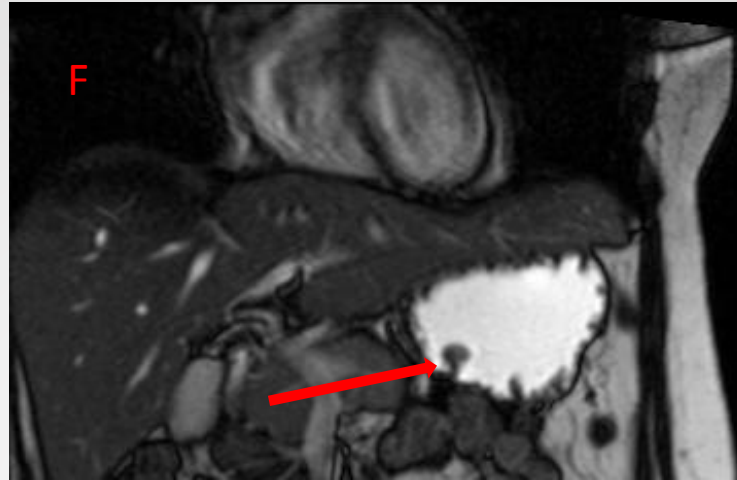
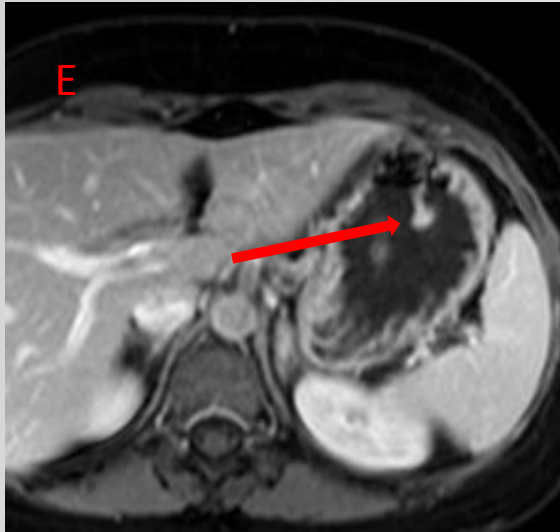


# Clinical Vignette No. 1

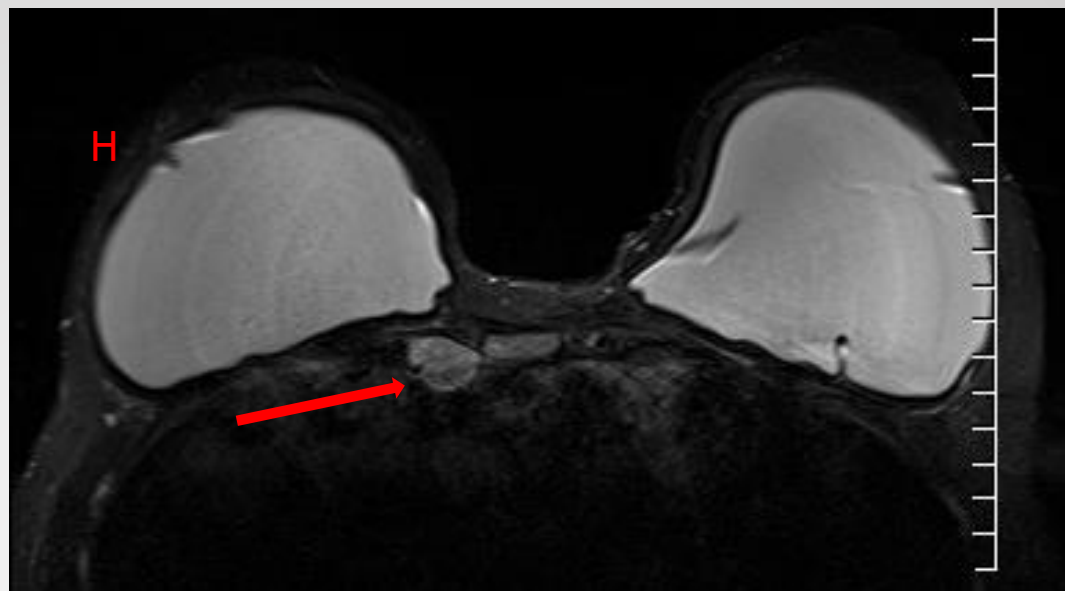
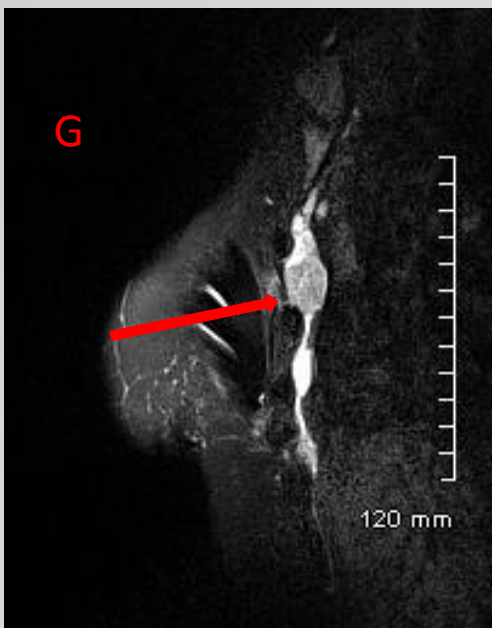
- 38 year old Female with PJS and multiple hamartomatous polyps throughout her GIT
- MRI and CT images (Image A,B,C, red arrows) demonstrate a chronic ileocecal intussusception with a 16 mm ileal luminal polyp as a lead point (image D, green arrow)



- MRI (Images E, F) demonstrate a pedunculated polyp in the body of the stomach (Red arrows)

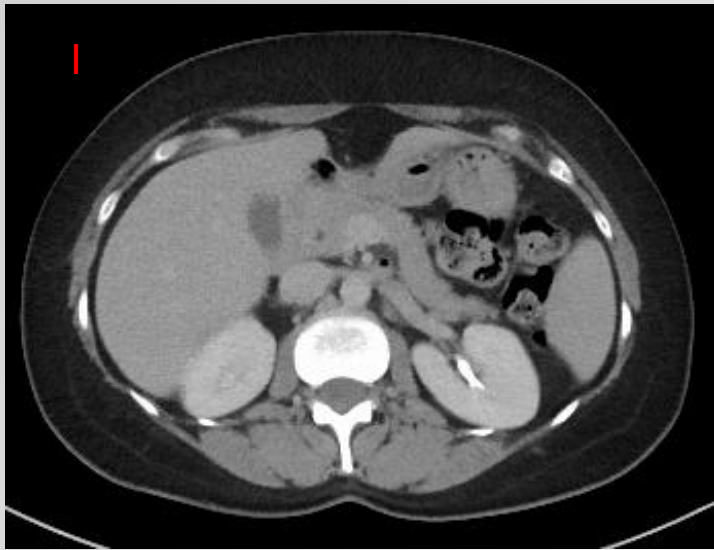


- Polyps occur in over 90% of individuals with PJS during their lifetime
- 50% of patients will experience intussusception or bowel obstruction before the age of 20
- GI bleeding is also a common complication.
- Overall, approximately 30% of the PJS patients required laparotomy before the age of 10 and 68% before the age of 18
- Regular small-bowel surveillance in PJS patients aims to reduce polyp-related complication, and to detect PJS associated bowel malignancy, although there is no data that supports the reduction in risk *via* surveillance[

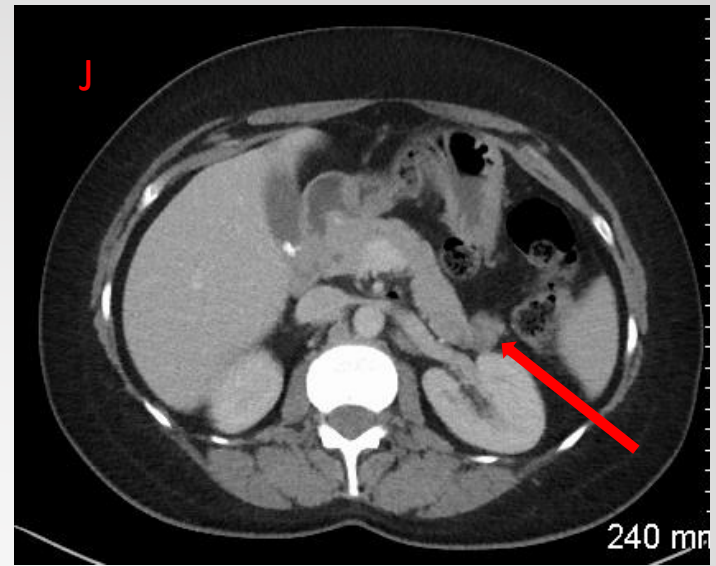


This same patient has a history of DCIS of the right breast in 2002 for which she underwent bilateral mastectomy (prophylactic on the left) and implant reconstruction. On annual surveillance MRI breast performed 2011, (Images G,H), she was noted to have recurrence in a right internal mammary lymph node (Red arrows)

- The cumulative breast cancer risk is estimated between 31%-54% at age 60, with a mean diagnosis age of 37
- The risk of breast cancer in PJS is similar to that of women with BRCA gene mutation
- There is no consensus with regards appropriate screening in this high risk patient cohort
- Giardiello recommends annual mammography or MRI starting at the age of 25 years
- Beggs et al suggest that annual MRI/ultrasound surveillance should start at age 25-30 years, substituted with mammography after the age of 50



May 2014



January 2017

This patient commenced chemotherapy for metastatic recurrent breast cancer and as a result entered into a CT surveillance program to monitor for response to treatment and disease progression.

Image I demonstrates a normal pancreatic body and tail. The next dedicated abdominal imaging study was not performed for 32 months, at which time, a CT study performed for investigation of abdominal pain (Image J) demonstrated an incidental 16mm pancreatic tail malignancy (red arrow)

- The risk of pancreatic cancer in these individuals is unclear, and varies between 7% and 36% by the age of 60