

# BC CRC Update

## Unusual Colorectal Tumors

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# Disclosures

- I have no disclosures

# Objectives

- Neuroendocrine Tumors (Carcinoids)
  - Appendiceal
  - Rectal
- Rectal GISTs
- Melanoma
  - Small bowel
  - Anorectal
- ColoRectal Lymphoma

# Quality of Evidence Poor!!!



# Neuroendocrine Tumors of Appendix

- Most common appendiceal tumor
- Most are found incidentally
  - 1 per 300 appendectomies
- Overall mets 4%, distant mets 0.7%
- Size predicts metastatic potential

# Size Matters!

- < 1cm
  - Rarely metastasize - Appy
- 1-2cm
  - Depends
- >2cm
  - Risk of lymph node mets 30% - right hemi

# High Risk Features for 1-2cm

- Invasion into mesoappendix
- Lymphovascular invasion
- Serosal involvement
- Involved margins
- Positive lymph nodes in appy specimen
- Hi Ki 67 index (>2%)
- Goblet cell variant

# Does Resection affect Survival?

- Most would probably assume so.
- SEER database – 576 pts (1988-2005)
  - Tumor size > 2cm predicted LN mets
  - No difference in survival between R hemi and appy



# Goblet Cell Carcinoid / Adenocarcinoid AKA Mixed endocrine / exocrine tumor

- Rare variant
- Aggressive
  - Often see peritoneal disease at presentation
- Treatment
  - Right hemicolectomy regardless of size, consideration of adj chemo, consideration of peritoneal stripping & HIPEC when disseminated
- Overall 10 year survival 60%

# Rectal Carcinoid (or NET of Rectum)

- Incidence increasing
  - 11% of GI NETSs & 1.5% of rectal neoplasms
  - 10 fold increase past 35 yrs
    - Scherubl. Endoscopy. 2009;41 (2):162-165
- More common in African descent
- Typically > 55 yrs of age
- Most discovered incidentally
- Prognosis size dependent

# Size Matters (again)

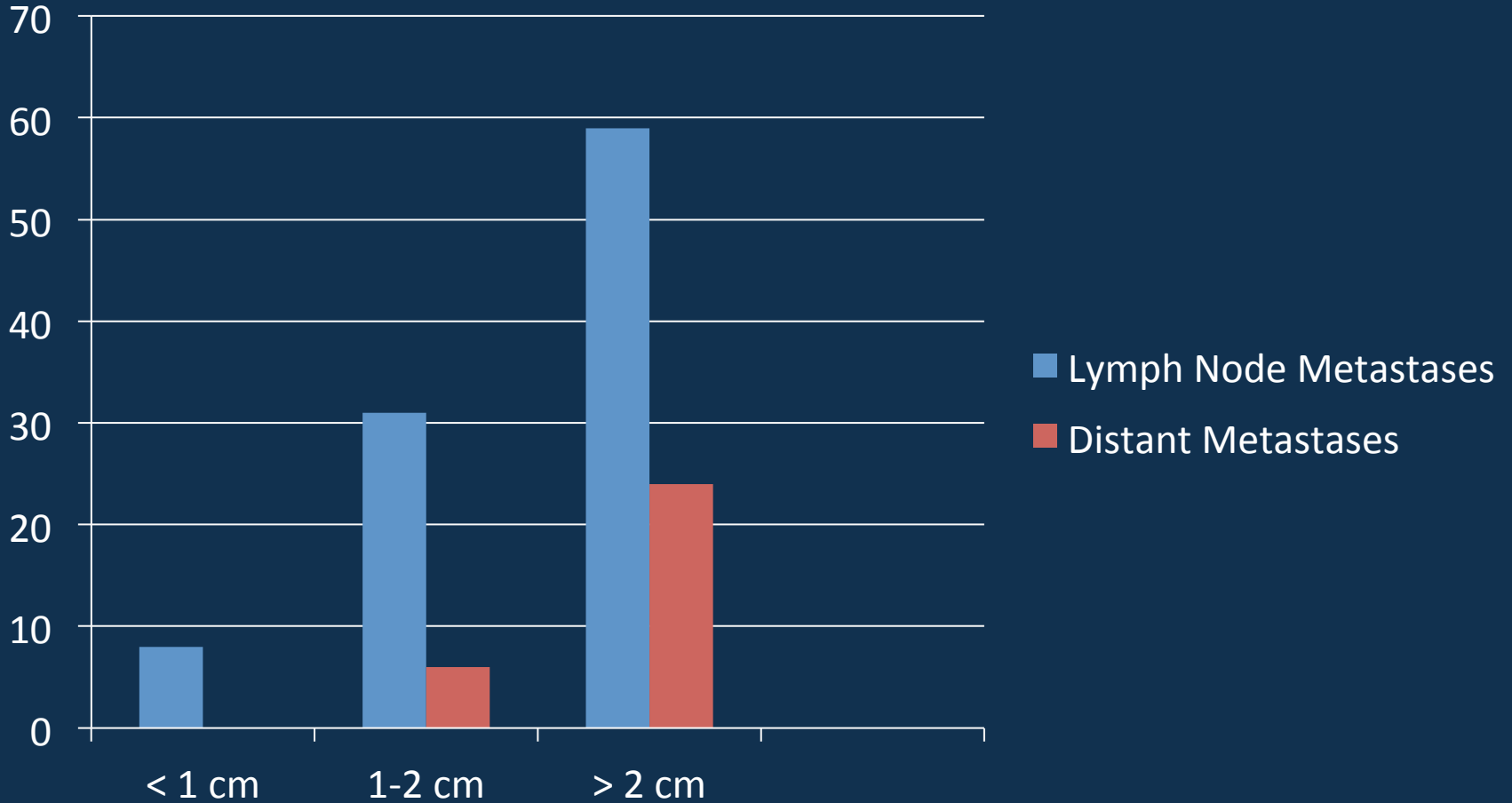
- < 1 cm
  - Incidence of LN involvement 0-3%
  - Excise endoscopically, TEM, or transanally
- 1-2 cm
  - Regional or distant disease 7-34%
  - Treatment controversial
- > 2 cm
  - Regional and distant spread 67%-100%
  - Radical excision indicated

**TABLE 2.** Rates of Nodal Involvement and Distant Metastases (n = 100)

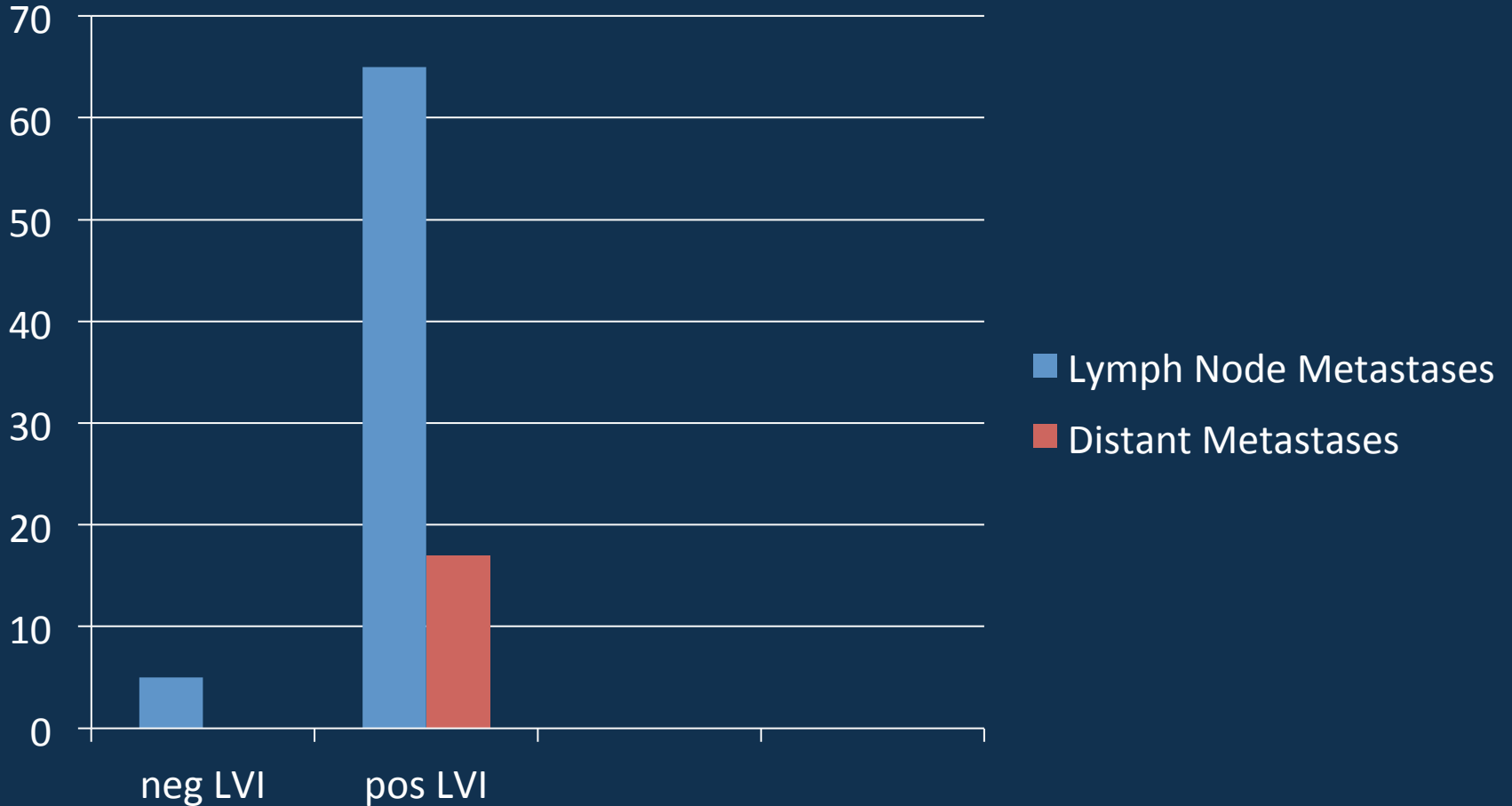
	<b>Nodal Involvement</b>	<b>Total, n (%)</b>	<b>P</b>	<b>Metastases</b>	<b>Total, n (%)</b>	<b>P</b>
<b>Sex</b>						
Male	19	62 (30%)	0.55	7	62 (11%)	0.489
Female	15	38 (38%)		5	38 (12.8%)	
<b>Age, y</b>						
≤60	19	54 (35%)	0.357	2	54 (4%)	<0.001
>60	15	46 (32%)		10	46 (21%)	
<b>Tumor size (mm)</b>						
1–10	2	25 (8%)	0.038	0	25 (0%)	0.049
11–20	11	36 (31%)		2	36 (6%)	
>21	17	29 (59%)		7	29 (24%)	
<b>Tumor depth</b>						
T1	4	32 (13%)	<0.001	0	32 (0%)	0.029
T2	8	25 (32%)		1	25 (4%)	
T3	14	29 (48%)		5	29 (17%)	
T4	6	11 (55%)		3	11 (27%)	
<b>Lymphovascular invasion</b>						
–ve	2	41 (5%)	< 0.001	0	38 (0%)	<0.001
+ve	24	37 (65%)		6	36 (17%)	
<b>Distance from anal verge, mm</b>						
≤70	10	39 (26%)	0.476	2	39 (5%)	0.046
≥71	17	47 (36%)		7	43 (16%)	
<b>Treatment</b>						
Ante	12	41 (29%)	0.271	4	41 (10%)	0.050
AR and TME	14	45 (31%)		2	45 (4%)	
Hartmann's procedure	3	7 (21%)		3	14 (21%)	
APR	3	7 (21%)				

APR indicates abdomino-perineal resection; AR, anterior resection; TME, total mesorectal excision.

# Effect of Size on Metastases



# Effect of LVI on Metastases



# Rectal GIST

- 7% of all GISTs
- Typically over 50 yrs
- Male predominance
- Frequent positive margins on excision (40%)
- Express KIT – so can be targeted with imatinib

# Rectal GIST

- Behaviour

- Based on size, Number of mitoses per 50 high power fields
- $< 2\text{cm}$  &  $< 5$  mitoses per 50 HPF
  - No risk of metastatic disease
- 2-5 cm &  $< 5$  mitoses per 50 HPF
  - Low risk of metastatic disease
- $> 5$  cm or any tumor size with  $> 5$  mitoses per HPF
  - High risk of metastatic disease ( $> 50\%$ )



# Rectal GIST - Approach

- Clearly resectable, acceptable morbidity
  - Primary Surgery
- Resectable but not without significant morbidity
  - Neoadjuvant imatinib
- Unresectable
  - Neoadjuvant or palliative imatinib

# Small Bowel Melanoma

- **Most** are metastatic lesions
- Difficult to be certain if primary or met
  - Criteria: No evidence of concurrent melanoma or atypical melanocytic lesion, absence of metastases other than regional nodes, presence of intramucosal lesion
- Worse prognosis than cutaneous melanoma

# Small Bowel Melanoma

- 60% of pts who die of melanoma have GI mets
  - Only 1.5%-4% are diagnosed
- Typically see multiple polypoid masses
- Present as other SB tumors do
- Surgery indicated for symptom control, esp if no evidence of disseminated disease
- Median survival 6-9 months, 15 months if completely resected
- 5 year survival < 10%

# AnoRectal Melanoma

- Very rare!
  - 0.2% of all melanoma
  - 0.1%-4.6% of all malignant tumors of the rectum & anus
  - Can be melanotic or amelanotic
  - Outcome very poor
    - 5 yr disease free survival 6.7%-12%
    - Median survival 19 months

# Anorectal Melanoma

## Local vs Radical Surgery

- Most reports indicate no difference in outcome between local & radical excision
  - » Nilsson & Ragnarsson-Olding. Br J Surg. 2010;97:98-103
  - » Kiran et.al. Dis Colon Rectum. 2010; 53: 402-408
  - » Homsy & Garrett. Dis Colon rectum. 2007; 50: 1004-1010
  - » Thibault et.al. Dis Colon Rectum. 1997; 40: 661-668
  - » Ross et al. Arch Surg. 1990;125: 313-316
- One case series (MSK) suggested advantage to APR (84 pts over 65 yr period, 71 not metastatic)
  - » Brady et.al. Dis Colon Rectum. 1995; 38: 145-151

# ColoRectal Lymphoma

- Again very rare!
  - 0.2-0.6% of colorectal malignancies
  - Dawson's criteria to establish primary CR lymphoma
  - In our series, rectal lymphoma was associated with reduced median survival (42 vs 110 months); and pts having surgical resection had improved survival (110 vs. 56 months)

Thanks!

# TABLE 3

**Carcinoid Tumors of the Rectum: A Multi-institutional International Collaboration.**

Shields, Conor; MD, FRCSI; Tiret, Emmanuel; Winter, Desmond; MD, FRCSI

Annals of Surgery. 252(5):750-755, November 2010.  
DOI: 10.1097/SLA.0b013e3181fb8df6

**TABLE 3. Multiple Variable Logistic Regression Analysis of Risk Factors for Lymph Node Metastases\***

<b>Variables</b>	<b>Odds Ratio</b>	<b>95% CI</b>	<b>P</b>
Tumor size > 10 mm	32.7	14.8–72.3	0.006
Lymphovascular invasion	19.6	12.3–146.0	< 0.001

\*Risk factors for nodal involvement calculated in patients who underwent formal surgical resection (n = 100).



# TABLE 4

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**TABLE 4. Multiple Variable Logistic Regression Analysis of Risk Factors for Distant Metastases\***

TABLE 4 . Multiple Variable Logistic Regression Analysis of Risk Factors for Distant Metastases\*

<b>Variables</b>	<b>Odds Ratio</b>	<b>95% CI</b>	<b>P</b>
Lymph node metastases	12.3	1.8-84.7	0.033
Lymphovascular invasion	74.4	4.6-120.2	0.022

\*Risk factors for distant metastases calculated in patients who underwent formal surgical resection (n = 100).