20210051

References:
#1: CAP Protocol for the Examination of Specimens From Patients With Carcinoma of the Ampulla of Vater. AmpullaVater 4.1.0.0, Feb 2020

#2: ICD-O-3, 49

Question:
Primary site/Biliary tract--Ampulla of Vater: What is the correct primary site code for intra-ampullary and periampullary adenocarcinoma, C241 (8144/3) or C249? See Discussion.

Discussion:
Ampulla, biopsy: High grade dysplasia with focal intramucosal carcinoma in a background of ulceration with acute and chronic inflammation.

Surgery pathology: Head of pancreas, duodenum, and distal stomach, pancreaticoduodenectomy-Ampulla, Adenocarcinoma, intestinal type, intra-ampullary and peri-ampullary (mixed type). Grade moderately differentiated, 1.5cm. Tumor invades into duodenal submucosa. Lymphovascular Invasion: Foci suspicious for lymphovascular invasion identified. Perineural Invasion: Present.

Synoptic report: Tumor Site Intra-ampullary and peri-ampullary (mixed type). Histologic Type Adenocarcinoma, intestinal type.

There is not enough information regarding site in radiology reports or operative report. CT-A/P/C: The patient's known ampullary mass is not well visualized on this exam. No significant intrahepatic or extrahepatic biliary ductal dilation is identified. The pancreatic duct is normal caliber.

Answer:
Assign C241. Ampulla (C241) includes both periampullary and intra-ampullary.

Date Last Modified:
10/01/2021

20210050

References:
2018 EOD Manual. General Instructions; Testis schema

Question:
EOD 2018/Extension--Testis: How is Extent of Disease (EOD) Primary Tumor coded if it appears limited to testis on scrotal ultrasound and is treated with neoadjuvant chemotherapy prior to the orchiectomy when there is no residual tumor (staged as ypT0 disease) and in cases where there is residual tumor? See Discussion.

Discussion:
Unless there is a biopsy that proves in situ tumor (EOD code 000, Tis) or extra testicular invasion into the scrotum, penis, or further contiguous extension (EOD code 700, T4), EOD Primary Tumor must be coded based on the PATHOLOGICAL assessment (orchiectomy). There are no other CLINICAL codes because the AJCC indicates imaging is not used for local T-categorization, and the EOD derives the AJCC TNM staging. If the case cannot be coded to either EOD Primary Tumor codes 000 or 700 clinically, the only clinical code that seems to apply is 999 (Unknown).

We are seeing more cases treated with neoadjuvant chemotherapy prior to orchiectomy, especially in patients with distant metastatic disease. The EOD Manual indicates that clinical evidence takes priority over pathological evidence when neoadjuvant treatment is given, unless the extent of disease following neoadjuvant treatment is greater than pre-treatment clinical findings. If the clinical and pathological information are the same, code the extension based on the clinical information.

Do these general rules also apply to testis even though we cannot code CLINICAL findings for these tumors? If so, will EOD Primary Tumor be coded to 999 (Unknown) for any testis primary that is not in situ or invasive into the scrotum, etc., that is treated with neoadjuvant therapy? Or should the post-neoadjuvant PATHOLOGICAL assessment be coded for these tumors because the CLINICAL assessment would otherwise be unknown?

How is the EOD Primary Tumor coded for the following two cases?

1. Left testicular mixed germ cell tumor, biopsy-proven metastasis to a supraclavicular lymph node. The left testis contained a small mass on scrotal ultrasound. The patient underwent neoadjuvant chemotherapy, and the post-treatment orchiectomy proved no residual primary tumor (ypT0). Is EOD Primary Tumor 999 because it is clinically unknown (even though it was clinically limited) or 800 (No evidence of primary tumor) because there was no pathological evidence of tumor following neoadjuvant treatment?

2. Right testicular mixed germ cell tumor with biopsy-proven inguinal lymph node metastasis. There was a palpable mass in right testis on physical exam (not described as fixed or involving scrotum). The patient underwent neoadjuvant chemotherapy, and the post-treatment orchiectomy proved a residual 2 cm tumor limited to the testis without lymphovascular invasion (LVI). Is EOD Primary Tumor 999 because it is clinically unknown, or 200 (PATHOLOGICAL assessment only - Limited to testis WITHOUT LVI)?

Answer:
Assign code 999 to EOD Primary Tumor for testis when neoadjuvant therapy is given, and clinical assignment is unknown, and the extent of the primary tumor is not fully assessed due to post neoadjuvant treatment effect as with the two case scenarios.

Both clinical examination and histologic (pathologic) confirmation are required by AJCC for clinical assessment and was not met in these scenarios.

While EOD Primary Tumor is based on pathologic assessment, the EOD general instructions are to code the clinical information if that is the farthest extension when the patient received neoadjuvant systemic therapy unless the post-neoadjuvant surgery shows more extensive disease. As there is neoadjuvant treatment effect and there is no clinical assessment, the primary tumor cannot be fully assessed.

Date Last Modified:
10/01/2021

20210049

References:
Heme & Lymphoid Manual and Database. September 2020; Effective with Cases Diagnosed 1/1/2010 and Forward

Question:
Histology/Heme & Lymphoid Neoplasms--Leukemia: Is this the correct histology for a case of acute myeloid leukemia (AML) with recurrent genetic abnormalities? If the only information was AML with recurrent genetic abnormalities, "what code would you use: AML, NOS (9861/3) or AML with recurrent genetic abnormalities (9896/3)? See Discussion.

Discussion:
12/3/2020 Pathology: AML: Blasts 40% of nucleated cells. CD45 positive, CD34 negative, CD 117+,

CD13 positive, CD33 positive in 59.6% and HLA-DR was dim, and myeloperoxidase was dim.

Cytogenetics normal karyotype. The next generation sequencing detected IDH 2p.(R172K)c515>A.

Because this was AML NOS, we consulted with the physician. The physician stated the patient had AML with recurrent genetic abnormalities and, the basis for the diagnosis was the IDH-2 mutation identified on Next Generation Sequencing. We assigned 9896/3, based on the physician's interpretation of the pathology. This histology is being questioned.

Answer:
We found that the term AML with recurrent genetic abnormalities, NOS was incorrectly included as an alternate name with code 9896/3. We followed back with our expert hematopathologist and he stated that this should have been coded to 9861/3 (AML, NOS), for AML with recurrent genetic abnormalities, NOS. This alternate name has been added to 9861/3. (Note: The same alternate name has been removed from 9896/3).

IDH-2 is not listed as a genetic abnormality for any of the histologies listed in the database. It could be that this is a new genetic marker for one of the AML with recurrent genetic abnormalities that we are not aware of. Without further clarification on which histology the IDH-2 would indicate, you would have to default to 9861/3.

There are several histologies that are grouped as AML with recurrent genetic abnormalities. All of these have specific genetics listed as part of the ICD-O-3 histology name.

9865: Acute myeloid leukemia with t(6;9) (p23;q34.1) DEK-NUP214

9866: Acute promyelocytic leukemia with PML-RARA

9869: Acute myeloid leukemia with inv(3)(q21.3q26.2) or t(3;3)(q21.3;q26.2); GATA2, MECOM

9871: Acute myeloid leukemia with inv(16)(p13.1q22) or t(16;16)(p13.1;q22); CBFB-MYH11

9877: Acute myeloid leukemia with mutated NPM1 (2021+)

9878: Acute myeloid leukemia with biallelic mutation of CEBPA (2021+)

9879: Acute myeloid leukemia with mutated RUNX1 (2021+)

9896: Acute myeloid leukemia with t(8;21)(q22;q22.1); RUNX1-RUNX1T1

9897: Acute myeloid leukemia with t(9;11)(p21.3;q23.3); KMT2A-MLLT3

9911: Acute myeloid leukemia (megakaryoblastic) with t(1;22)(p13.3;q13.1); RBM15-MKL1

9912: Acute myeloid leukemia with BCR-ABL1 (2021)+

Of note, for the above histologies, since these are diagnosed solely based on genetics, diagnostic confirmation will always be 3. This instruction will be added to the Hematopoietic database for the 2022 update.

Date Last Modified:
10/01/2021

20210048

References:
#1: ICD-O-3.2

#2: WHO Class Digestive System Tumors, 202-204. Anal squamous dysplasia

Question:
Reportability--Anal Canal: Is a 2021 diagnosis of moderate squamous dysplasia (AIN II) of the anal canal reportable? See Discussion.

Discussion:
We are aware that squamous intraepithelial neoplasia, grade II (e.g., AIN II), 8077/2 is reportable for 2021. However, because this is also called rather than high grade squamous dysplasia (8077/2), we are unsure about reportability. There is no known histology and behavior code for moderate squamous dysplasia, the classifications available are only low grade (8077/0) or high grade (8077/2).

Answer:
If possible, clarify with the pathologist/physician what is meant by "moderate squamous dysplasia (AIN II)."

If no further information can be obtained, report this case based on the diagnosis of "AIN II." Squamous intraepithelial neoplasia, grade II is listed in ICD-O-3.2 as 8077/2 making it reportable for cases diagnosed in 2021. AIN is a type of squamous intraepithelial neoplasia.

Date Last Modified:
08/12/2021

20210047

References:
#1: Summary Stage 2018. v2.0; Digestive System Sites

#2: SEER*RSA. Colon and Rectum, Extent of Disease Primary Tumor

Question:
Summary Stage 2018/EOD 2018--Colon: Does the 2018 SEER Summary Staging Manual, Digestive System Sites, Distinguishing In Situ and Localized Tumors for the Digestive System, #1. b., Exception, include in situ plus intramucosal carcinoma (involvement of the lamina propria and may involve but not penetrate through the muscularis mucosa) (penetration through the muscularis mucosa is behavior code 3.)? This seems to be in conflict with Extent of Disease (EOD) 2018. See Discussion.

Discussion:
We are preparing to send our hospitals a reminder that the behavior changes from 2 to 3 at the bottom of the basement membrane, and the T category changes from Tis to T1 at the bottom of the mucosa for colon and rectum carcinomas. We are confused by the wording of the Exception.

Distinguishing In Situ and Localized Tumors for the Digestive System:
1.b. If the tumor has penetrated the basement membrane to invade the lamina propria, in which case it is localized and assigned Summary Stage 1 (localized) and for invasion of the lamina propria

Exception: Code 0 (behavior code 2) includes cancer cells confined within the glandular basement membrane (intraepithelial); includes in situ plus intramucosal carcinoma (involvement of the lamina propria and may involve but not penetrate through the muscularis mucosa) (penetration through the muscularis mucosa is behavior code 3.)

The text following (intraepithelial) is unclear. The question is: Does the text include in situ plus intramucosal carcinoma (involvement of the lamina propria and may involve but not penetrate through the muscularis mucosa) (penetration through the muscularis mucosa is behavior code 3.) mean the following:

Code 0 (behavior code 2) includes in situ plus intramucosal carcinoma. In situ plus intramucosal carcinoma is involvement of the lamina propria, which may involve (but not penetrate through) the muscularis mucosae. Penetration through the muscularis mucosa is behavior 3. If that is what the text above means, then it seems that the 2018 SEER Summary

Stage Manual is saying colorectal tumors reported as: adenocarcinoma in situ, at least intramucosal adenocarcinoma in situ, high grade dysplasia/intramucosal adenocarcinoma in situ, focally intramucosal at the margin are to be coded behavior 2 and SEER Summary stage In situ (0) like the intraepithelial carcinoma tumors. However, it conflicts with the EOD Data for Colon and Rectum, Note 2, and SINQ 20210006. The text for both EOD Data for Colon and Rectum and SINQ 20210006 is clear. According to them, the above bulleted adenocarcarcinoma examples are coded SEER Summary Stage localized (1) and behavior 3. SINQ 20210006 states that: For purposes of Summary Stage, intramucosal carcinoma is a localized lesion So, intramucosal carcinoma is coded SEER Summary Stage 1 (localized) and (behavior code 3).

According to the text for EOD Primary Tumor, Colon and Rectum, Note 2 below, intramucosal, NOS involvement is invasive.
Note 2: Code 050 (behavior code 3) includes the following:

Intramucosal, NOS

Lamina propria

Mucosa, NOS

Confined to, but not through the muscularis mucosa

Thank you for your help clarifying the 2018 SEER Summary Manual Exception text above.

Answer:
For purposes of Summary Stage, intramucosal, NOS is a localized lesion. Intramucosal carcinoma is coded SEER Summary Stage 1 (localized) and (behavior code 3). The involvement of the following is assigned localized in Summary Stage and assigned a behavior code of 3.

Intramucosal, NOS

Lamina propria

Mucosa, NOS

Confined to, but not through the muscularis mucosa

The Exception you cite may need to be reworded. We will review for the next version of the Summary Stage manual.

Date Last Modified:
08/05/2021

20210046

References:
#1: WHO Class Skin Tumors, 304-305. 4th edition

#2: ICD-O-3.2

Question:
Reportability--Skin: Is dermatofibrosarcoma protuberans (DFSP) with fibrosarcomatous transformation synonymous with dermatofibrosarcoma protuberans, fibrosarcomatous, and therefore reportable for diagnosis year 2021 and forward? See Discussion.

Discussion:
Patient has a 2021 skin excision showing an atypical spindle cell neoplasm, most consistent with dermatofibrosarcoma protuberans (DFSP) with fibrosarcomatous transformation.

Per the ICD-O-3.2 Coding Table, DFSP, NOS has a behavior code of /1, and DFSP, fibrosarcomatous has a behavior code of /3. There is no code listed for DFSP with fibrosarcomatous transformation. Transformation is not included as a term that can/cannot be used for the Other Sites Schema, but this type of DFSP is often described as DFSP with fibrosarcomatous transformation. How do we code DFSP when transformation is used to describe fibrosarcomatous?

Answer:
Report DFSP with fibrosarcomatous transformation as it is synonymous with fibrosarcomatous DFSP (8832/3). According to the WHO Classification of Skin Tumors, 4th edition, fibrosarcomatous DFSP is a variant of DFSP and that fibrosarcomatous transformation is seen in approximately 10% of DFSP cases. It is characterized by an often-abrupt transition of DFSP.

Date Last Modified:
08/02/2021

20210043

References:
#1: 2021 SEER Manual, 6. Reportability

#2: WHO Classification of Tumors Female Genital Tract. 5th edition; online version

Question:
Reportability--Fallopian Tube: Is a diagnosis of serous tubal intraepithelial neoplasm (neoplasia) (STIN) equivalent to serous tubal intraepithelial carcinoma (STIC)? Does the designation of high or low grade have any effect on potential reportability? See Discussion.

Discussion:
Patient has left salpingo-oophorectomy showing fallopian tube with focal high grade serous intraepithelial neoplasm.

In reviewing some journal articles, the term STIN is being used to describe both STIC and serous tubal intraepithelial lesion (STIL). We will likely continue to see this term used, so it would be nice to have some clarity.

Answer:
Serous tubal intraepithelial neoplasm (neoplasia) (STIN) is **not** equivalent to serous tubal intraepithelial carcinoma (STIC). Report STIN only when stated to be high grade. STIC is reportable. Do not report STIL.

According to our expert pathologist consultant, STIL and STIN are broad descriptive terms that reflect proliferation of epithelial cells with varying degrees of atypia, with the most developed, STIC, reflecting convincing neoplastic change.

Date Last Modified:
08/03/2021