



UNION EUROPÉENNE DES MÉDECINS SPÉCIALISTES EUROPEAN UNION OF MEDICAL SPECIALISTS

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Syllabus for residents and trainees in Rare Adult Solid Cancers

The basic goal of this syllabus is to provide an understanding between the instructor and trainee so there is minimal confusion in the topics, with clear expectations. It is not a classical syllabus as it contains descriptions from different areas, but it still summarizes major and specific topics that should be covered during the training course of a resident. This syllabus is intended as supporting reference material, and the precise content and priorities of training may vary in different training institutions. The syllabus can also be modified to reflect each instructor's teaching philosophy towards the trainees.

1. There are scientific publications, web pages, and conference materials available online that could be used for educational purposes for various types of rare adult solid cancers. This is a comprehensive summary of them.
2. There are significant differences in the number of available scientific publications and reviews for different rare adult solid cancers. Some, like sarcomas, have a very robust literature, while others have been sparsely researched and consequently the availability of study materials is quite poor.
3. These differences also apply to life events and natural history. In the list of the EU CE accredited events there is a strong underrepresentation for some types of rare adult solid cancers.
4. Some conferences in this area have a long history, and the thought leaders in the specific fields are involved. In such cases the agendas of the conferences are designed to provide excellent education about best clinical practices for these rare adult solid cancers, opportunities to share major advances in research, and sessions that support the development of new collaborations and new investigators. For other cancers established conferences with solid reputations do not yet exist.
5. These imbalances will persist unless policy makers and research funders provide more attention for research, treatment, and networking on underrepresented rare tumour types. They may also be addressed by the ERNs, or by the UEMS, as recognised stakeholders in improving medical education.
6. As well, there are barriers in the communication of JARC proposals to the ERNs. All ERNs that are involved in rare cancer have made very impressive progress in all fields, including

education. However, communication among ERNs about their education efforts is sparse and not officially regulated. Therefore, we will not have in the near future mutual and harmonised indicators for successful knowledge implementation, which will hinder efforts at assessment.

Domain 1.: Fields of rare adult solid cancers/literature

1.1. Head and neck cancers

1.1.1. Epithelial tumours of nasal cavity and sinuses

Qin Y, Lu Y, Zheng L, Liu H. Ghost cell odontogenic carcinoma with suspected cholesterol granuloma of the maxillary sinus in a patient treated with combined modality therapy: A case report and the review of literature. *Medicine*. 2018;97(7):e9816. Epub 2018/02/15. doi: <https://doi.org/10.1097/md.0000000000009816>. PubMed PMID: 29443742; PubMed Central PMCID: PMC5839843.

Takakura H, Tachino H, Fujisaka M, Nakajima T, Yamagishi K, Ishida M, et al. Lymphoepithelial carcinoma of the maxillary sinus: A case report and review of the literature. *Medicine*. 2018;97(28):e11371. Epub 2018/07/12. doi: <https://doi.org/10.1097/md.0000000000011371>. PubMed PMID: 29995775; PubMed Central PMCID: PMC6076030.

Thompson LDR, Franchi A. New tumor entities in the 4th edition of the World Health Organization classification of head and neck tumors: Nasal cavity, paranasal sinuses and skull base. *Virchows Archiv : an international journal of pathology*. 2018;472(3):315-30. Epub 2017/04/27. doi: <https://doi.org/10.1007/s00428-017-2116-Q>. PubMed PMID: 28444451.

Zhang N, Zhou B, Huang Q, Chen X, Cui S, Huang Z, et al. Multiple metastases of clear-cell renal cell carcinoma to different region of the nasal cavity and paranasal sinus 3 times successively: A case report and literature review. *Medicine*. 2018;97(14):e0286. Epub 2018/04/06. doi: <https://doi.org/10.1097/md.0000000000010286>. PubMed PMID: 29620646; PubMed Central PMCID: PMC5902287.

Zhenwei C, Zhaoming W, Hongqi S, Qinwei L. Renal cell -like carcinoma of the nasal cavity: a case report and review of the literature. *Diagnostic pathology*. 2017;12(1):75. Epub 2017/10/19. doi: <https://doi.org/10.1186/s13000-017-0660-1>. PubMed PMID: 29041930; PubMed Central PMCID: PMC5645912.

1.1.1.1. Squamous cell carcinoma with variants of nasal cavity and sinuses

1.1.1.2. Lymphoepithelial carcinoma of nasal cavity and sinuses

1.1.1.3. Undifferentiated carcinoma of nasal cavity and sinuses

1.1.1.4. Intestinal type adenocarcinoma of nasal cavity and sinuses

1.1.2. Epithelial tumours of nasopharynx

Outh-Gauer S, Alt M, Le Tourneau C, Augustin J, Broudin C, Gasne C, et al. Immunotherapy in head and neck cancers: A new challenge for immunologists, pathologists and clinicians. *Cancer treatment reviews*. 2018;65:54-64. Epub 2018/03/17. doi: <https://doi.org/10.1016/j.ctrv.2018.02.008>. PubMed PMID: 29547766.

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Zhang WL, Ma S, Havrilla L, Cai L, Yu CQ, Shen S, et al. Primary thyroid-like low-grade nasopharyngeal papillary adenocarcinoma: A case report and literature review. *Medicine*. 2017;96(47):e8851. Epub 2018/02/01. doi: <https://doi.org/10.1097/md.00000000000008851>. PubMed PMID: 29381996; PubMed Central PMCID: PMC5708995.

Sun Q, Chen M, Sun Y, Chen X, Xu H, Rong L, et al. Cervical metastasis of gingival carcinoma misdiagnosed as branchiogenic carcinoma, a rare entity - report of a case and review of literature. *BMC oral health*. 2017;17(1):139. Epub 2017/12/01. doi: <https://doi.org/10.1186/s12903-017-0435-9>. PubMed PMID: 29183323; PubMed Central PMCID: PMC5706288.

1.1.2.1. Squamous cell carcinoma with variants of nasopharynx

1.1.2.2. Papillary adenocarcinoma of nasopharynx

1.1.3. Epithelial tumours of major salivary glands and salivary-gland type tumours

Li Q, Huang P, Zheng C, Wang J, Ge M. Prognostic significance of p53 immunohistochemical expression in adenoid cystic carcinoma of the salivary glands: a meta-analysis. *Oncotarget*. 2017;8(17):29458-73. Epub 2017/02/17. doi: <https://doi.org/10.18632/oncotarget.15297>. PubMed PMID: 28206977; PubMed Central PMCID: PMC5438744.

Lopez F, Williams MD, Skalova A, Hellquist H, Suarez C, Nixon IJ, et al. How Phenotype Guides Management of the Most Common Malignant Salivary Neoplasms of the Larynx? *Advances in therapy*. 2017;34(4):813-25. Epub 2017/02/23. doi: <https://doi.org/10.1007/s12325-017-0494-y>. PubMed PMID: 28224460.

Ord RA, Ghazali N. Margin Analysis: Malignant Salivary Gland Neoplasms of the Head and Neck. *Oral and maxillofacial surgery clinics of North America*. 2017;29(3):315-24. Epub 2017/05/30. doi: <https://doi.org/10.1016/j.j.coms.2017.03.008>. PubMed PMID: 28551337.

Falk N, Weissferdt A, Kalhor N, Moran CA. Primary Pulmonary Salivary Gland-type Tumors: A Review and Update. *Advances in anatomic pathology*. 2016;23(1):13-23. Epub 2015/12/10. doi: <https://doi.org/10.1097/pap.0000000000000099>. PubMed PMID: 26645458.

Cuthbertson DW, Raol N, Hicks J, Green L, Parke R. Minor salivary gland basal cell adenocarcinoma: a systematic review and report of a new case. *JAMA otolaryngology—head & neck surgery*. 2015;141(3):276-83. Epub 2015/01/03. doi: <https://doi.org/10.1001/jamaoto.2014.3344>. PubMed PMID: 25555241.

1.1.3.1. Epithelial tumours of major salivary glands

1.1.3.2. Salivary gland type tumours of head and neck

1.1.4. Epithelial tumours of hypopharynx and larynx

Caroppo D, Salerno G, Merolla F, Mesolella M, Ilardi G, Pagliuca F, et al. Coexistent Squamous Cell Carcinoma and Granular Cell Tumor of Head and Neck Region: Report of Two Very Rare Cases and Review of the Literature. *International journal of surgical pathology*. 2018;26(1):47-51. Epub 2017/08/09. doi: <https://doi.org/10.1177/1066896917724513>. PubMed PMID: 28783989.

Zhang Q, Xu H, You Y, Zhang J, Chen R. High Gpx1 expression predicts poor survival in laryngeal squamous cell carcinoma. *Auris, nasus, larynx*. 2018;45(1):13-9. Epub 2017/06/24. doi: <https://doi.org/10.1016/j.anl.2017.05.012>. PubMed PMID: 28641905.

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Wollenberg B. Cancer Immunology and HPV. Recent results in cancer research *Fortschritte der Krebsforschung Progres dans les recherches sur le cancer*. 2017;206:243-8. Epub 2016/10/05. doi: https://doi.org/10.1007/978-3-319-43580-0_19. PubMed PMID: 27699544.

1.1.4.1. Squamous cell carcinoma with variants of hypopharynx

1.1.4.2. Squamous cell carcinoma with variants of larynx

1.1.5. Epithelial tumours of oropharynx

Panwar A, Interval E, Lydiatt WM. Emergence of a Novel Staging System for Oropharyngeal Squamous Cell Carcinoma Based on HPV Status. *Oncology (Williston Park, NY)*. 2017;31(12):e33-e40. Epub 2018/01/04. PubMed PMID: 29297174.

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1.1.5.1. Squamous cell carcinoma with variants of oropharynx

1.1.6. Epithelial tumours of oral cavity and lip

Madera Anaya MV, Franco JV, Merchan-Galvis AM, Gallardo CR, Bonfill Cosp X. Quality assessment of clinical practice guidelines on treatments for oral cancer. *Cancer treatment reviews*. 2018;65:47-53. Epub 2018/03/17. doi: <https://doi.org/10.1016/j.ctrv.2018.03.001>. PubMed PMID: 29547765.

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Zhang WB, Peng X. Cervical metastases of oral maxillary squamous cell carcinoma: A systematic review and meta-analysis. *Head & neck*. 2016;38 Suppl 1:E2335-42. Epub 2016/02/19. doi: <https://doi.org/10.1002/hed.24274>. PubMed PMID: 26890607.

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1.1.6.1. Squamous cell carcinoma with variants of oral cavity

1.1.6.2. Squamous cell carcinoma with variants of lip

1.1.7. Epithelial tumours of eye and adnexa

Ford J, Thakar S, Thuro B, Esmali B. Prognostic Value of the Staging System for Eyelid Tumors in the 7th Edition of the American Joint Committee on Cancer Staging Manual. *Ophthalmic plastic and reconstructive surgery*. 2017;33(5):317-24. Epub 2017/03/30. doi: <https://doi.org/10.1097/iop.0000000000000901>. PubMed PMID: 28350693.

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Viani GA, Fendi LI. Adjuvant treatment or primary topical monotherapy for ocular surface squamous neoplasia: a systematic review. *Arquivos brasileiros de oftalmologia*. 2017;80(2):131-6. Epub 2017/06/08. doi: <https://doi.org/10.5935/0004-2749.20170032>. PubMed PMID: 28591290.

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1.1.7.1. Squamous cell carcinoma with variants of eye and adnexa

1.1.7.2. Adenocarcinoma with variants of eye and adnexa

1.1.8. Epithelial tumours of middle ear

Cugley DR, Roberts-Thomson SJ, McNab AA, Pick Z. Biopsy-Proven Metastatic Merkel Cell Carcinoma to the Orbit: Case Report and Review of Literature. *Ophthalmic plastic and reconstructive surgery*. 2018;34(3):e86-e8. Epub 2018/03/06. doi: <https://doi.org/10.1097/iop.0000000000001078>. PubMed PMID: 29505467.

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Yang Y, Zhou J, Wu H. Diagnostic value of sentinel lymph node biopsy for cT1/T2N0 tongue squamous cell carcinoma: a meta-analysis. *European archives of oto-rhino-laryngology : official journal of the European Federation of Oto-Rhino-Laryngological Societies (EUFOS) : affiliated with the German Society for Oto-Rhino-Laryngology - Head and Neck Surgery*. 2017;274(11):3843-52. Epub 2017/09/14. doi: <https://doi.org/10.1007/s00405-017-4740-3>. PubMed PMID: 28900723.

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1.1.8.1. Squamous cell carcinoma with variants middle ear

1.1.8.2. Adenocarcinoma with variants of middle ear

1.2. Thoracic rare cancers

1.2.1. Epithelial tumour of trachea

1.2.1.1. Squamous cell carcinoma with variants of trachea

Moores D, Mane P. Pathology of Primary Tracheobronchial Malignancies Other than Adenoid Cystic Carcinomas. Thoracic surgery clinics. 2018;28(2):149-54. Epub 2018/04/09. doi: <https://doi.org/10.1016/j.thorsurg.2018.01.003>. PubMed PMID: 29627048.

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1.2.1.2. Adenocarcinoma with variants of trachea

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Varela P, Pio L, Torre M. Primary tracheobronchial tumors in children. Seminars in pediatric surgery. 2016;25(3):150-5. Epub 2016/06/16. doi: <https://doi.org/10.1053/j.sempedsurg.2016.02.013>. PubMed PMID: 27301601.

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1.2.1.3. Salivary gland type tumours of trachea

Moores D, Mane P. Pathology of Primary Tracheobronchial Malignancies Other than Adenoid Cystic Carcinomas. Thoracic surgery clinics. 2018;28(2):149-54. Epub 2018/04/09. doi: <https://doi.org/10.1016/j.thorsurg.2018.01.003>. PubMed PMID: 29627048.

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1.2.2. Rare epithelial tumours of lung

1.2.2.1. Adenosquamous carcinoma of lung

Guo LC, Li G, Wang XM, Zhang M, Huang JA, Chen YB. Penile metastases from primary lung cancer: Case report and literature review. Medicine. 2017;96(26):e7307. Epub 2017/06/29. doi: <https://doi.org/10.1097/md.00000000000007307>. PubMed PMID: 28658136; PubMed Central PMCID: PMC5500058.

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1.2.2.2 Large cell carcinoma of lung

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1.2.2.3. Salivary gland type tumours of lung

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1.3. Male genital rare cancers

1.3.1. Rare epithelial tumours of prostate

1.3.1.1. Squamous cell carcinoma with variants of prostate

1.3.1.2. Infiltrating duct carcinoma of prostate

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1.3.1.4. Salivary gland type tumours of prostate

1.3.2. Testicular and paratesticular cancers

1.3.2.1. Paratesticular adenocarcinoma with variants

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1.3.3.1. Squamous cell carcinoma with variants of penis

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1.3.3.2. Adenocarcinoma with variants of penis

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1.3.8.1. Non seminomatous germ cell tumours

1.3.8.2. Seminomatous germ cell tumors

1.3.8.3. Germ cell tumors of central nervous system (CNS)

1.4. Female genital rare cancers

1.4.1. Rare epithelial tumours of breast

1.4.1.1. Mammary paget's disease of breast

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1.4.2. Rare epithelial tumours of corpus uteri

1.4.2.1. Squamous cell carcinoma with variants of corpus uteri

1.4.2.2. Adenoid cystic carcinoma of corpus uteri

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1.4.2.5. Mullerian mixed tumour of corpus uteri

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1.4.3.1. Squamous cell carcinoma with variants of cervix uteri

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1.4.3.3. Undifferentiated carcinoma of cervix uteri

1.4.3.4. Mullerian mixed tumour of cervix uteri

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1.4.4. Epithelial tumours of ovary and fallopian tube

1.4.4.1. Adenocarcinoma with variants of ovary

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1.4.4.2. Mucinous adenocarcinoma of ovary

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1.4.4.3. Clear cell adenocarcinoma of ovary

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1.4.4.5. Mullerian mixed tumour of ovary

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1.4.4.6. Adenocarcinoma with variant of fallopian tube

1.4.5. Non epithelial tumours of ovary

1.4.5.1. Sex cord tumours of ovary

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1.4.5.3. Germ cell tumour of ovary

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1.4.6. Epithelial tumours of vulva and vagina

1.4.6.1. Squamous cell carcinoma with variants of vulva and vagina

1.4.6.2. Adenocarcinoma with variants of vulva and vagina

1.4.6.3. Paget's disease of vulva and vagina

1.4.6.4. Undifferentiated carcinoma of vulva and vagina

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1.4.7. Trophoblastic tumour of placenta

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1.5 Urological / Urogenital rare cancers

1.5.1. Rare epithelial tumours of kidney

1.5.1.1. Squamous cell carcinoma spindle cell type of kidney

1.5.1.2. Squamous cell carcinoma with variants of kidney

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1.5.2. Epithelial tumours of pelvis and ureter

1.5.2.1. Transitional cell carcinoma of pelvis and ureter

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1.5.2.2. Squamous cell carcinoma with variants of pelvis and ureter

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1.5.2.3. Adenocarcinoma with variants of pelvis and ureter

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1.5.3. Epithelial tumours of urethra

1.5.3.1. Transitional cell carcinoma of urethra

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1.5.3.2. Squamous cell carcinoma with variants of urethra

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1.5.3.3. Adenocarcinoma with variants of urethra

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1.5.4. Rare epithelial tumours of bladder

1.5.4.1. Squamous cell carcinoma with variants of bladder

1.5.4.2. Adenocarcinoma with variants of bladder

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1.5.4.3. Salivary gland type tumours of bladder

1.6. Neuroendocrine tumours

1.6.1. Rare neuroendocrine tumours

1.6.1.1. GEP - well differentiated not functioning endocrine carcinoma of pancreas and digestive system

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1.6.1.7. Typical and atypical carcinoid of the lung

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1.6.1.8. Rare neuroendocrine carcinoma of other sites

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1.10.1.2. Adenocarcinoma with variants of oesophagus

1.10.1.3. Salivary gland type tumours of oesophagus

1.10.1.4. Undifferentiated carcinoma of oesophagus

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1.10.8.4. Adenocarcinoma with variants of liver and IBT

1.10.8.5. Undifferentiated carcinoma of liver and IBT

1.10.8.6. Squamous cell carcinoma with variants of liver and IBT

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1.10.9.1. Adenocarcinoma with variants of gallbladder

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1.10.9.3. Squamous cell carcinoma of gallbladder and EBT

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Domain 2.: Special web materials

The following links provide valuable, comprehensive informations and/or educational materials occasionally with clinico-pathological consequences about almost all cancer types including rare variants. Not only may they be useful for health care professionals and researchers, graduate students and postgraduate physician but also for patients and their caregivers or their relatives. Furthermore you may see the following books related to this topic, with well-formed illustration of the cancer types, about the macroscopy and their histological morphology. They also give information about the most important differential diagnosis, including the differences between the common and rare variants as well. According to our experience in case of rare diseases (especially in cases of such malignancies) it's important to include them into the differential.

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2.1. Links

<http://www.pathologyoutlines.com/>
<http://www.webpathology.com>
<http://uscap.sclivelearningcenter.com/Index.aspx?PID=2870>
<http://knowledgehub.uscap.org/index.htm?hub.htm>
<http://apps.pathology.jhu.edu/sp/>
<http://www.uab.edu/medicine/pathology/education/cases>
<http://path.upmc.edu/casemonth/ap-casemonth.html>
<http://www.virtualpathology.leeds.ac.uk/cow/cow.php?year=2013>
<https://med.nyu.edu/pathology/caseoftheweek>
<https://medicine.hsc.wvu.edu/pathology/case-of-the-month/>
<https://www.oncolink.org/>
<https://www.cancer.gov/>
<http://www.cancerindex.org/>

2.2. Books

Textbook of Uncommon Cancer, 5th Edition
Manual of Clinical Oncology, 7th Edition
Pediatric Oncology: A Comprehensive Guide, 3rd Edition
Lanzkowsky's Manual of Pediatric Hematology and Oncology, 6th Edition
Oncology Boards Flash Review, 1st Edition
New Trends in Cancer for the 21st Century, 1st Edition
Targeted Therapies for Solid Tumors: A Handbook for Moving Toward New Frontiers in Cancer Treatment
Successes and Limitations of Targeted Cancer Therapy
Melanoma: Translational Research and Emerging Therapies, 1st Edition
Cancer of the Head and Neck, 5th Edition
Series WHO:
WHO Classification of Tumours of Central Nervous System. Revised 4th edition
WHO Classification of Tumours of Haematopoietic and Lymphoid Tissues. Revised Fourth Edition
WHO Classification of Tumours of the Digestive System. Fourth Edition
WHO Classification of Tumours of the Breast. Fourth Edition
WHO Classification of Tumours of Soft Tissue and Bone. Fourth Edition
WHO Classification of Tumours of Female Reproductive Organs. Fourth Edition
WHO Classification of Tumours of Lung Pleura, Thymus and Heart. Fourth edition
WHO Classification of Tumours of the Urinary System and Male Genital Organs. Fourth edition
WHO Classification of Head and Neck Tumours. Fourth edition
WHO Classification of Tumours of Endocrine Organs. Fourth Edition
Pathology and Genetics of Tumours of the Skin. Third edition
Series: Diagnostic Pathology
Diagnostic Pathology: Pediatric Neoplasms

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Diagnostic Pathology: Endocrine
Diagnostic Pathology: Blood and Bone Marrow
Diagnostic Pathology: Lymph Nodes and Extranodal Lymphomas
Diagnostic Pathology: Bone
Diagnostic Pathology: Thoracic
Diagnostic Pathology: Hepatobiliary and Pancreas
Diagnostic Pathology: Neoplastic Dermatopathology
Diagnostic Pathology: Genitourinary
Diagnostic Pathology: Head and Neck
Diagnostic Pathology: Neuropathology
Diagnostic Pathology: Breast
Diagnostic Pathology: Vascular
Diagnostic Pathology: Molecular Oncology
Diagnostic Pathology: Kidney Diseases
Diagnostic Pathology: Soft Tissue Tumors
Diagnostic Pathology: Gastrointestinal
Diagnostic Pathology: Gynecological
Diagnostic Pathology: Placenta
Diagnostic Pathology: Familial Cancer Syndromes