

# PEDIATRIC PATHOLOGY WARM-UP QUESTIONS

**1. An ovarian tumor from a young child is most likely to belong to which GENERAL category?**

- A. Germ cell tumor
- B. Sex cord stromal tumor
- C. Epithelial tumor
- D. Metastasis

**2. Which of the following is NOT a classic “small round blue cell tumor?”**

- A. Neuroblastoma
- B. Ewing sarcoma
- C. PNET
- D. Desmoplastic small round cell tumor
- E. Triphasic Wilm’s Tumor
- F. CLL/SLL

**3. True or false: Prognostic rules for adult tumors can be applied to pediatric tumors of the same histologic type.**

**4. A 4-year-old child presents with a pathologic fracture of the femur. Imaging reveals multiple distinct bone lesions as well as a large adrenal-based mass. On histology the tumor is comprised of small round blue cells. What is the most likely diagnosis?**

- A. Neuroblastoma
- B. Wilm’s tumor
- C. Pheochromocytoma
- D. Germ cell tumor
- E. Ewing sarcoma

**5. The most common renal neoplasm in *newborn* babies is:**

- A. Congenital mesoblastic nephroma
- B. Wilm’s tumor
- C. Rhabdoid tumor
- D. Clear cell sarcoma
- E. Angiomyolipoma

# PEDIATRIC PATHOLOGY CHALLENGE QUESTIONS

**1. All of the following are features of monophasic synovial sarcoma except:**

- A. PAS-positive secretions
- B. Hemangiopericytoma-like vascular pattern
- C. Microvilli on EM
- D. SYT fusion gene
- E. Epithelioid cell clusters highlighted by cytokeratin 7 and 19

**2. Which of the following variants of synovial sarcoma is associated with a favorable prognosis?**

- A. Biphasic
- B. Monophasic
- C. Calcified
- D. Poorly-differentiated monophasic
- E. SYT-SSX1 gene fusion

**3. A 20-year-old man with a family history of autoimmune disease presents with subcutaneous nodules involving his wrist and elbow. Histology demonstrates palisaded nodules with central necrosis and a robust inflammatory infiltrate. What study *must* be performed?**

- A. Serum rheumatoid factor
- B. Cytokeratin immunostain
- C. Polarization to look for birefringence patterns
- D. GMS stain

**4. You receive a slide from a painful subungual mass showing epithelioid cells bearing round, regular nuclei surrounded by eosinophilic cytoplasm. Which of the following positive stains would confirm your diagnosis?**

- A. smooth muscle actin
- B. cytokeratin 5/6
- C. CD34
- D. CD31
- E. Desmin

**5. A 12-year-old girl presents with a painless 2 cm hand mass. Histology shows a fibrous background containing cellular nodules of primitive mesenchymal cells surrounding central**

**calcification and chondroid tissue. The fibrous component is cytologically bland but infiltrates adjacent muscle, fat, and nerve. This suggests a diagnosis of:**

- A. Epithelioid sarcoma
- B. Calcifying aponeurotic fibroma
- C. Chondrosarcoma
- D. Fibrosarcoma

**6. Excisional biopsy of a cheek lesion from a 10-year-old reveals an admixture of macrophages, bland spindled cells, and occasional large cells bearing a core of eosinophilic cytoplasm surrounded by a wreath of nuclei and rimmed by lipid vacuoles. We would expect all of the following immunostains to be positive except:**

- A. CD68
- B. vimentin
- C. lysozyme
- D. factor XIIIa
- E. cytokeratin

**7. All of the following are features of alveolar rhabdomyosarcoma except:**

- A. More common in adolescents than young children
- B. Worse prognosis than embryonal rhabdomyosarcoma
- C. Tumor cells are arranged around alveoli-like spaces lined by fibrous septa
- D. Most common location is head and neck
- E. Multinucleated giant cells with a wreath of nuclei are highly suggestive of this rhabdomyosarcoma subtype

**8. Which single statement about embryonal rhabdomyosarcoma is correct?**

- A. Cases in children have a worse prognosis than cases in adolescents
- B. Cross-striations are readily identifiable
- C. Paratesticular and extremity tumors are most common in young children
- D. Classic histology shows a myxoid background with a dense cambium layer underlying the epithelium
- E. Cells usually have eccentric nuclei and eosinophilic granular cytoplasm

**9. A 15-year-old girl with a history of hundreds of colon polyps and appendectomy presents with an abdominal mass. Histology shows a uniformly cellular tumor comprised of cytologically bland fibroblasts which merge with the background collagen. Staining for beta-catenin shows nuclear positivity. What other tumor might this patient be at risk for?**

- A. Jaw osteoma

- B. Benign colorectal polyps with prominent smooth muscle
- C. Endometrial carcinoma
- D. Cerebellar medulloblastoma
- E. Sebaceous carcinoma

**10. Infantile fibrosarcoma has the following translocation, shared with which other tumor:**

- A. t(X;18), synovial sarcoma
- B. t(12;15), cellular congenital mesoblastic nephroma
- C. t(17;22), dermatofibrosarcoma protuberans
- D. t(11;22), Ewings sarcoma
- E. t(15;19), NUT Midline carcinoma

**11. Which feature would not be expected in a fibrous hamartoma of infancy?**

- A. Fibrous trabecular
- B. Mature Fat
- C. Mature cartilage
- D. Immature mesenchymal cells
- E. Squamous hyperplasia of adjacent skin

**12. All of the following are features of inclusion body fibromatosis except:**

- A. Inclusions stain for PAS
- B. Dorsal digital nodules in patients under 1 year of age
- C. Inclusions are perinuclear and approximately the size of erythrocytes
- D. Spindle cells set in a collagen matrix
- E. Infiltration of the deep dermis is common

**13. A young woman presents with a polypoid vascular proliferation of the oral mucosa. The lesion is comprised of a lobular proliferation of small vessels lined by plump endothelial cells with a fibrous connective tissue background. Which of the following statements is true about this entity?**

- A. Tumor cells recapitulate the arterial component of a specialized arteriovenous shunt
- B. Distinctive features include “promontory sign,” hyaline globules, and plasma cells
- C. Associated with POEMS syndrome
- D. Clonal evidence suggest that they are true neoplasms
- E. Occurs at the result of thrombosis within preexisting vascular lesion

**14. Which of the following is a deep dermal/subcutaneous tumor seen predominantly in children/young adult females which is comprised of fibrohistiocytic cells and generic multinucleated giant cell with an associated fibrous rim?**

- A. Juvenile xanthogranuloma
- B. Plexiform fibrohistiocytic tumor
- C. Giant cell tumor of tendon sheath
- D. Benign fibrous histiocytoma

**15. A 15-year-old male presents with femur tumor centered in the epiphysis with metaphyseal extension. Histology demonstrates a proliferation of grooved mononuclear cells with eosinophilic cytoplasm and distinct cytoplasmic borders. "Chicken wire" calcification, scattered giant cells, and focal chondroid differentiation are present. This tumor is:**

- A. Osteosarcoma
- B. Chondroblastoma
- C. Osteoblastoma
- D. Non-ossifying fibroma
- E. Osteoid osteoma

**16. A 15-year-old male presents with a 2 cm femur tumor centered in the diaphysis. Imaging shows a radiolucent nidus surrounded by a sclerotic rim. Gross inspection reveals a red, granular region corresponding with the nidus identified on imaging. Histology shows anastomosing trabecula of woven bone with osteoblastic rimming with capillary proliferations between trabecula. This tumor is:**

- A. Osteosarcoma
- B. Fibrous dysplasia
- C. Osteoblastoma
- D. Non-ossifying fibroma
- E. Osteoid osteoma

**17. A 15-year-old male presents with a radiolucent, scalloped cortical defect of the femur centered on the metaphysis. Histology demonstrates spindle cells arranged in storiform patterns with frequent mitotic figures and admixed benign giant cells. This tumor is:**

- A. Osteosarcoma
- B. Chondroblastoma
- C. Osteoblastoma
- D. Non-ossifying fibroma
- E. Osteoid osteoma

**18. A 15-year-old male presents with a femur mass. Imaging reveals a destructive lesion centered on the metaphysis with cortical reactive bone and lifting of the adjacent periosteum. Histology shows an atypical spindle cell proliferation with a lacelike osteoid matrix formation and scattered benign giant cells.**

- A. Osteosarcoma
- B. Chondroblastoma
- C. Osteoblastoma
- D. Non-ossifying fibroma
- E. Osteoid osteoma

**19. A 12-year-old girl presents with a jaw mass with radiographically ill-defined borders. Histology shows a hypocellular proliferation of spindled cells with an “alphabet soup” of small woven bony trabecula. What syndrome is this patient at risk for?**

- A. Gardener’s Syndrome
- B. Albright Syndrome
- C. Cowden Syndrome
- D. Carney Complex

**20. All of the following statements about dermatofibrosarcoma protuberans are true except:**

- A. Bears the t(17;22) translocation
- B. Bednar tumor is a pigmented variant that must be distinguished from melanoma
- C. Infiltration into the dermis is common
- D. Metastasis is common
- E. Strongly CD34 positive

**21. All of the following are features of nodular tenosynovitis except:**

- A. Most common in hands
- B. Local recurrence is uncommon (20%)
- C. Florid papillary synovitis is present
- D. Histology shows cellular tumor with benign multinucleated giant cells, inflammatory cells, and hemosiderin
- E. COL6A3-CSF1 gene fusion described

**22. A 20-year-old male presents with a well-circumscribed tumor of the index finger. The surgeon reports that the tumor is closely associated with a tendon. On histology the lesion is densely collagenized and paucicellular with bland spindled cells bearing uniform elongate nuclei. Scattered slit-like vascular spaces are present. This tumor is:**

- A. Fibroma of tendon sheath
- B. Nodular tenosynovitis
- C. Inclusion body Fibromatosis
- D. Calcifying aponeurotic fibroma

**23. Microscopic examination of sections from a skin ellipse from a 12-year-old demonstrates a lens-shaped zone bearing spindled cells and thick collagen bundles. The overlying epithelium shows stromal induction. Foamy macrophages, hemorrhage, and Touton-like giant cells are characteristic of what variant of this tumor?**

- A. Deep penetrating
- B. Lipidized
- C. Aneurysmal
- D. Fibrotic
- E. Granular cell

**24. Schiller-Duval bodies are:**

- A. Eosinophilic fluid-filled spaces surrounded by a ring of tumor cells
- B. Papillae with central core containing a vessel
- C. Papillae associated rounded cores of calcium
- D. Eosinophilic, acellular areas surrounded by palisaded tumor cells
- E. Fibrillary eosinophilic material surrounded by a ring of tumor cells

**25. The most common pattern seen in ovarian yolk sac tumors is:**

- A. Hepatoid
- B. Microcystic
- C. Solid
- D. Polyvesicular vitelline
- E. Reticular
- F. Papillary

**26. The presence of scattered syncytiotrophoblastic cells in an otherwise embryonal-appearing ovarian tumor suggests a mixed germ cell tumor with a choriocarcinoma contribution. T/F?**

**27. All of the following are true of ovarian choriocarcinomas except:**

- A. High propensity for metastasis, especially to lungs
- B. Extremely chemoresponsive with cures occurring even after wide metastasis
- C. Proliferation of syncytiotrophoblasts with or without cytotrophoblasts

- D. Dilated vessels and hemorrhage are common features
- E. Accounts for a ~1% of ovarian germ cell tumors

**28. The presence of peritoneal gliomatosis portends aggressive disease progression in immature teratomas. T/F.**

**29. A 17-year-old woman presents with a right adnexal mass, elevated lactate dehydrogenase, and mildly elevated beta-HCG. Histology of the tumor reveals a diffuse arrangement of homogeneous tumor cells with faintly eosinophilic cytoplasm, prominent nucleoli, and well-demarcated cell membranes. Scattered syncytiotrophoblasts are appreciated. The background stroma is fibrous and a dense lymphocytic infiltrate peppers the tumor. The diagnosis is:**

- A. Mixed germ cell tumor
- B. Sertoli-Leydig cell tumor
- C. Yolk Sac tumor
- D. Dysgerminoma
- E. Seminoma
- F. Embryonal carcinoma

**30. All of the following are common features of juvenile granulosa cell tumor except (pick 2!):**

- A. May occur at any age
- B. Abundant eosinophilic or vacuolated cytoplasm
- C. Hyperchromatic atypical nuclei with prominent nucleoli
- D. Variably sized follicles containing mucicarmine-+ secretions
- E. Nuclear grooves
- F. Usually present at high stage

**31. You receive an adnexal mass from an 18-year-old girl. The requisition sheet notes that the patient has marked virilization. Histology reveals a tumor comprised of cells with vacuolated cytoplasm arranged in solid tubules with admixed nests of polyhedral cells bearing eosinophilic cytoplasm, eccentrically-located nuclei, and prominent nucleoli. Despite careful search, intracytoplasmic crystals are not identified. In one field, heterologous mucinous elements are noted. The diagnosis is:**

- A. Embryonal carcinoma
- B. Mixed germ cell tumor
- C. Sertoli-Leydig cell tumor
- D. Yolk sac tumor
- E. Mature teratoma



**32. A 13-year-old boy presents with a large femur mass and no evidence of additional masses elsewhere by imaging. Histology reveals a homogeneous proliferation of small round cells with finely stippled nuclei. Presuming that this tumor is primary to the bone, we could also see all of the following features except:**

- A. Intermediate filaments on electron microscopy
- B. t(21;22)
- C. Fibrillary eosinophilic material surrounded by a ring of tumor cells
- D. CD99 immunoreactivity
- E. Fibrous septa intersecting through groups of tumor cells
- F. PASd positivity
- G. All are correct

**33. A 19-year-old male has a large abdominal mass with nests of small round to spindled blue cells set in a prominent desmoplastic stroma. Given the histologic picture, tumor location, and the patient's age and gender you immediately suspect desmoplastic small round cell tumor. Which additional test result would help to seal the deal?**

- A. t(11;22) identical to what is seen in Ewings/PNET
- B. Dual positivity for cytokeratin and desmin
- C. Dual positivity for WT1 and myogenin
- D. Loss of INI expression
- E. Strong CD99 immunoeexpression

**34. A young child is brought in for a large abdominal mass. Imaging localizes the tumor to the adrenal gland and histology shows a small round blue cell proliferation with thin intersecting fibrovascular cores and scattered Homer Wright pseudorosettes. Unfavorable features could include all of the following except:**

- A. Stroma-rich with a nodular pattern
- B. Low mitotic karyorrhectic index (MKI)
- C. Differentiated tumor in a patient >60 months
- D. Undifferentiated tumor in a patient <18 months

**35. Which of the following patients with neuroblastomas falls into a "high risk" pretreatment group? (When not mentioned, assume molecular tests are negative)**

- A. A 4-year-old boy with non-metastatic intermixed ganglioneuroblastoma and MYCN amplification
- B. A 4-year-old boy with metastatic differentiating neuroblastoma
- C. A 1-year-old boy with metastatic poorly-differentiated neuroblastoma

- D. A 1-year-old boy with metastatic poorly-differentiated neuroblastoma and hyperdiploidy
- E. A 4-year-old boy with non-metastatic poorly-differentiated neuroblastoma with 11q aberrations

**36. A 3-year-old child presents with a renal mass that appears pale blue and encapsulated at low power. Higher power review reveals polygonal tumor cells divided into vague nests by fine vascular septa. The cell borders are imperceptible and chromatin is fine. Mucopolysaccharide vacuoles are present. This patient is most likely to have:**

- A. Organ-confined disease
- B. Widespread metastasis involving bone
- C. Metastases limited to lung, liver, and lymph nodes
- D. Bilateral tumors

**37. A 3-year old child presents with a renal mass comprised of blastemal, stromal, and epithelial elements. All of the following are true of this tumor except (pick 2):**

- A. May secrete Epo, renin, and Von Willebrand factor
- B. Anaplasia is identified in 5% of tumors and corresponds with p53 expression
- C. Nephrogenic rests indicate unfavorable histology
- D. Metastases most commonly involve bone and brain
- E. Most often occurs in children aged 2-5 years
- F. Loss of heterozygosity at 1p and 16q are associated with worse prognosis

**38. A hypercalcemic infant presents with a renal mass and a concurrent posterior fossa mass. The renal tumor is most likely to have:**

- A. Loss of INI1 expression
- B. Inactivation of chromosome 22
- C. Cytoplasmic inclusions containing intermediate filaments
- D. Large rounded cells with abundant cytoplasm and prominent nucleoli
- E. All of the above

**39. An incidentally discovered pediatric renal tumor comprised of infiltrative thin spindled cells forming “onionskin” rings around native renal elements with associated cartilage formation is most likely:**

- A. Wilms tumor
- B. A malignant relative of Wilms tumor
- C. A benign relative of Wilms tumor
- D. Congenital mesoblastic nephroma

**40. All of the following apply to pediatric adrenal cortical neoplasms except:**

- A. Typically positive for calretinin, inhibinA, and D240
- B. Carcinomas are associated with Li-Fraumeni syndrome
- C. Weiss histologic criteria are helpful for distinguishing carcinomas from adenomas
- D. Size >500 grams suggests malignancy
- E. Patients may present with virilization

**41. Skin biopsy from a 1-year-old baby shows a dermal proliferation of CD1a-positive cells with eosinophilic cytoplasm and folded, grooved, and reniform nuclei. Prominent admixed inflammatory component is appreciated. If the skin lesions are multifocal and systemic involvement is present, a diagnosis of what disease is warranted?**

- A. Letterer-Siwe disease
- B. Hand-Schüller-Christian disease
- C. Eosinophilic granuloma
- D. Histiocytosis X

**42. Which of the following statements about hepatoblastoma patterns is false?**

- A. Fetal pattern shows uniform cells which are larger than normal hepatocytes
- B. Fetal pattern has a “light and dark” appearance on low power due to variations in lipid and glycogen content
- C. Embryonal pattern has a component of pleomorphic angulated cells with little cytoplasm
- D. Macrotrabecular pattern has trabeculae >10 cells thick
- E. Extramedullary hematopoiesis is common in fetal and embryonal patterns

**43. An infant with Beckwith-Wiedemann syndrome presents with a primary pancreatic tumor. We could expect to see all of the following features except:**

- A. Squamoid nests
- B. Islands of polygonal epithelial cells separated by cellular stroma
- C. Acinar-type cells containing zymogen granules
- D. Immunostaining for synaptophysin and NSE
- E. Immunostaining for CEA and EMA
- F. All of the above

**44. Lymph node from a 10-year-old boy reveals large pleomorphic lymphocytes with CD30 positivity, frequent mitoses, and scattered cells bearing kidney-shaped nuclei with a perinuclear eosinophilic region. The proliferation infiltrates the sinuses and paracortex with relative follicular sparing. Which of the following statements is true?**

- A. t(2;5) translocation imparts a worsened prognosis

- B. ALK1 immunostaining is seen in a minority of cases
- C. ALK1 nuclear and cytoplasmic staining suggests a variant translocation
- D. Most cases show a T-cell phenotype
- E. Hallmark cells are a feature

**45. Which of the following statements about the molecular genetics of Burkitt lymphoma is false?**

- A. The classic translocation results in juxtaposition of myc and Ig heavy chain
- B.  $t(8;14)(q24;32)$  is seen in ~80%
- C. Variant translocations may involve 2p11 and 22q11
- D. The classic translocation is unique to Burkitt lymphoma
- E. All of the above are true

**46. An 18-month old boy presents with a primary testicular tumor. Without even seeing the slide, you bet your morning coffee that he has a:**

- A. Pure seminoma
- B. Mature teratoma
- C. Immature teratoma
- D. Yolk sac tumor
- E. Mixed germ cell tumor
- F. Spermatocytic seminoma

**47. The pediatric surgeon sends you another testicular tumor. Having retained your morning coffee in your last successful wager, you decide to risk it all again, this time betting that the tumor most certainly is not:**

- A. Pure seminoma
- B. Mature teratoma
- C. Immature teratoma
- D. Yolk sac tumor
- E. Mixed germ cell tumor
- F. Spermatocytic seminoma

**48. Which of the following is not a feature of Meckel Diverticulum?**

- A. Located 2 cm from ileocecal valve
- B. Present in 2% of population
- C. Ectopic pancreatic tissue common
- D. Represents persistence of the omphalomesenteric duct
- E. Outpouching of antimesenteric border of the terminal ileum

**49. All of the following statements about the plasma cell variant of Castleman disease are true except:**

- A. Associated with HIV
- B. Associated with HHV8
- C. Plasma cells are polytypic
- D. Classic finding is involuted follicular centers with penetrating hyalinized vessels
- E. Usually multicentric

**50. Ultrasound performed on a woman with polyhydramnios reveals a fetal upper lung lobe mass comprised of a dominant large cyst with several smaller associated cysts. This imaging finding is most consistent with which type of CCAM?**

- A. Type I
- B. Type II
- C. Type III
- D. Type IV