Unusual Cancer Sites

Annual Meeting

FCDS

Sept 10, 2026

BETTY MALANOWSKI

Prenatal origins of cancer

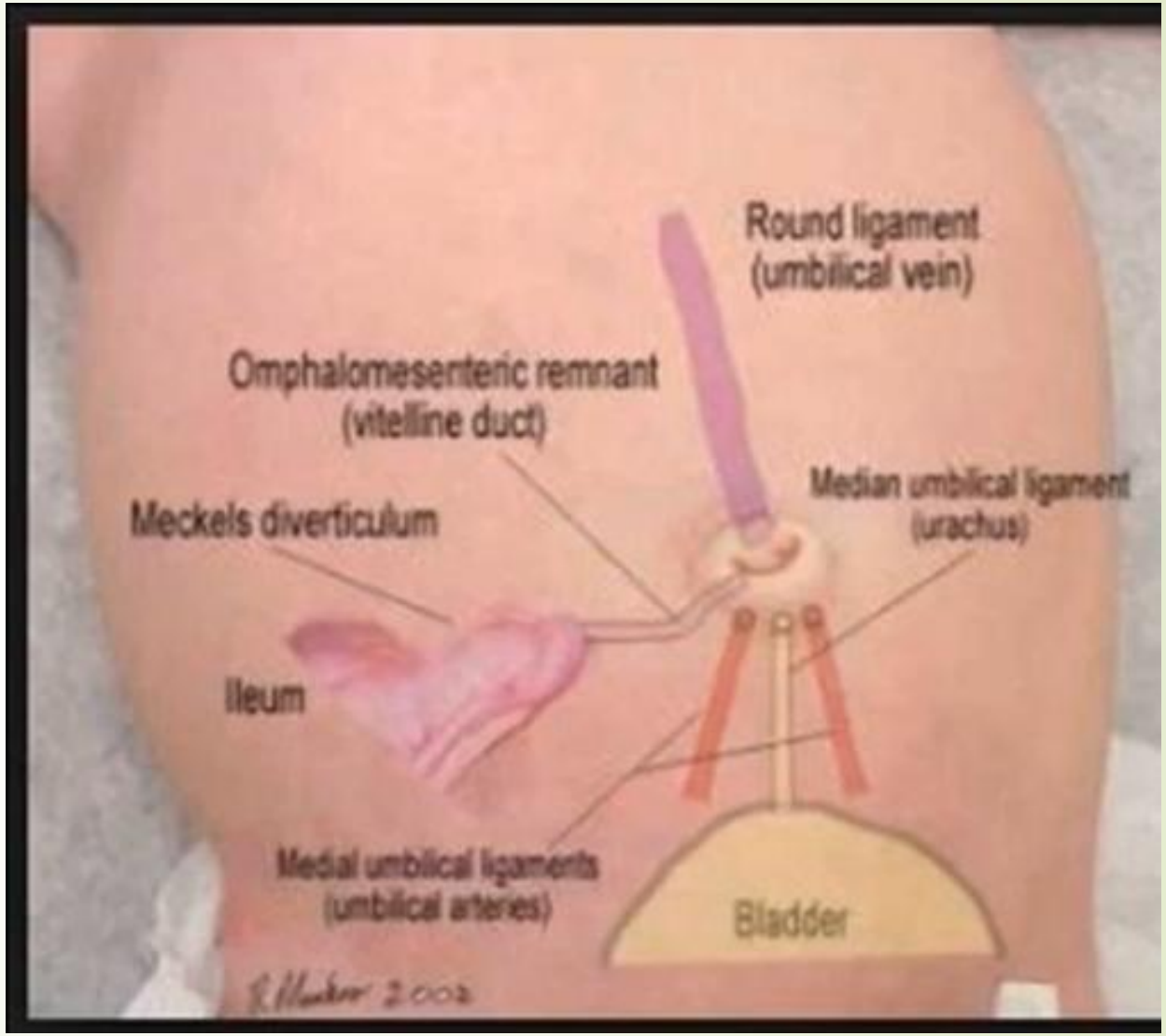
- Embryonal and Vestigial Structures

Sometimes we can get cancer from some remnant structures formed during the gestational period.

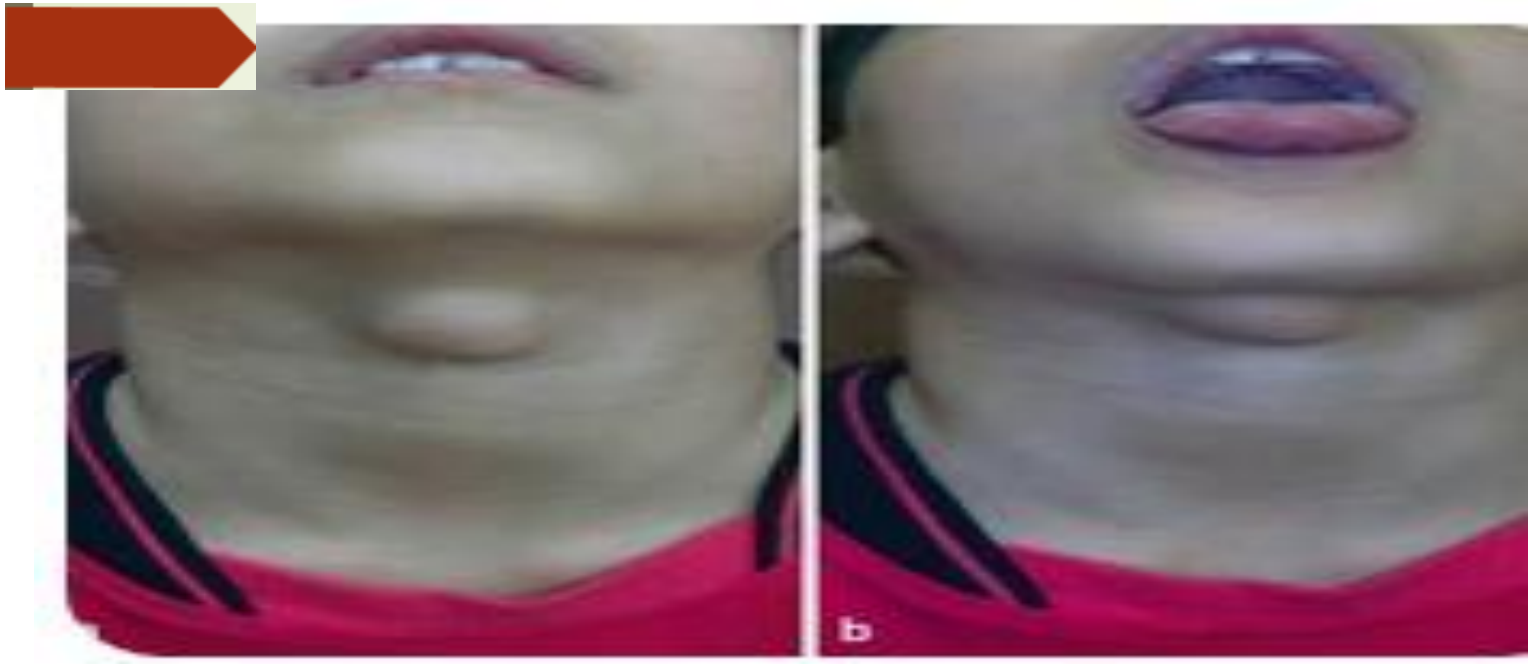
- Some of these structures include:

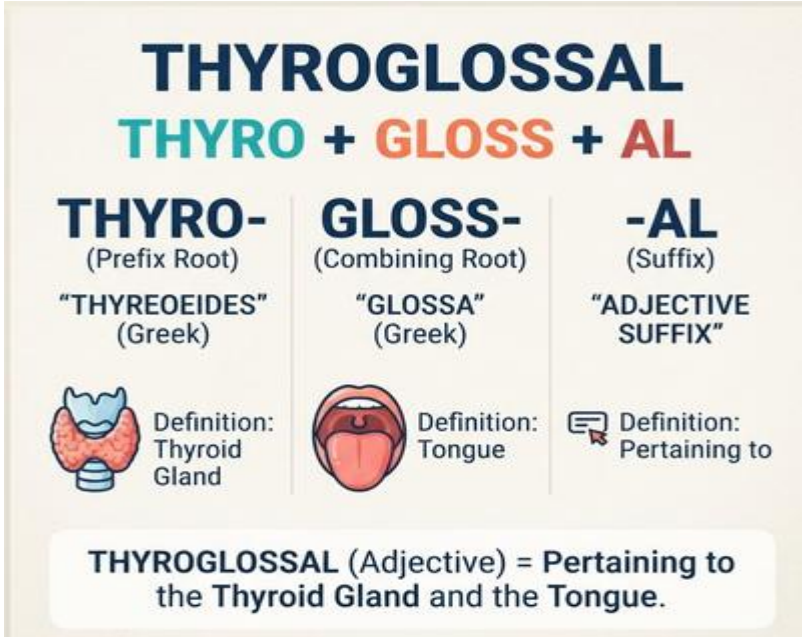
Primitive germ-cells remnants, residual neuroectodermal cells, embryonic gut or duct remnants.

- Some of these embryonical structures are supposed to disappear completely while others transform into vestigial structures.



Thyroglossal Duct Cyst Primary Site





CYST definition: sack-like structures filled with liquid, semisolid or gaseous material.

Code	Description
1	Thyroid gland Thyroid, NOS
2	Thyroglossal duct cyst

Thyroglossal duct

Temporary embryological tract that guides the developing thyroid gland from the base of the tongue (foramen cecum) to its final position in the neck.

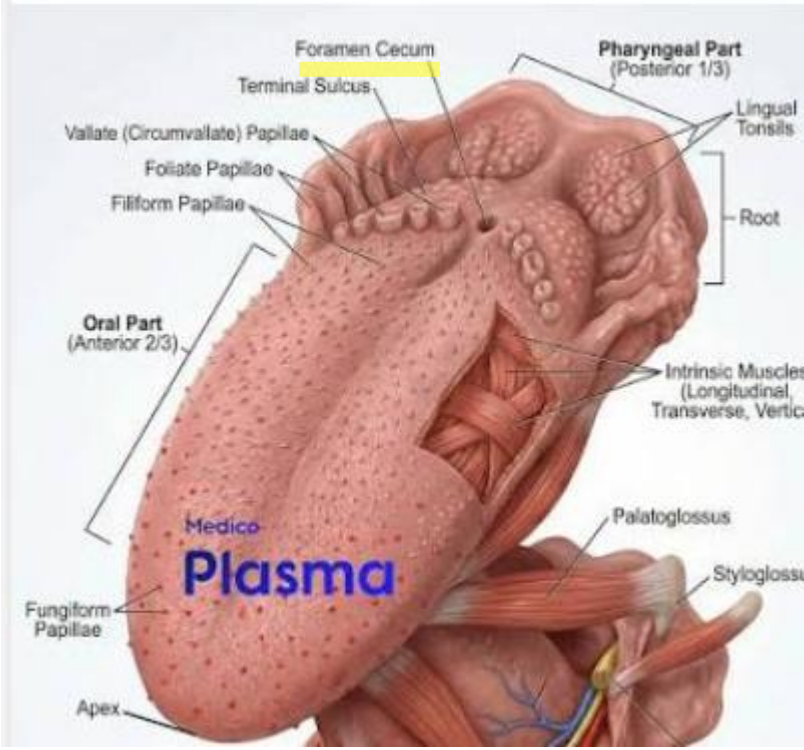
The **cells in the base of the tongue will form the thyroid gland**, but they must travel to its definitive location, so they travel down the neck through this temporary thyroglossal duct.

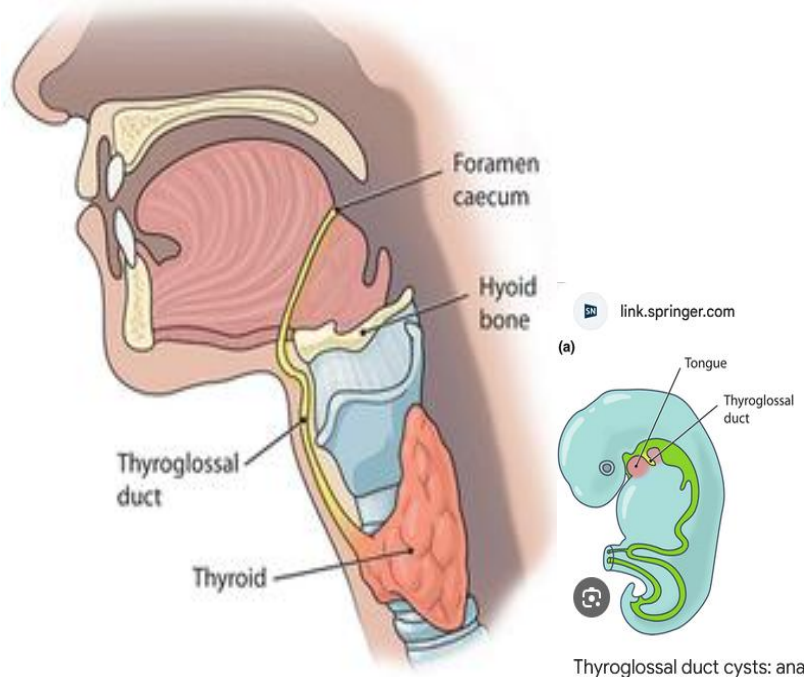
The cells reach the lower neck by the 8th to 10th week of pregnancy.

Once the journey ends, the duct is not needed anymore, and the thyroglossal duct disappears.

Sometimes parts of the duct or tunnel **fail to disappear**, and those leftover sections can fill with fluid, creating a **cyst**.

Thyroglossal duct cysts are benign, but those embryological remnant can be as site of malignancy!





Thyroglossal duct cyst and CANCER

- They are mostly benign**! 1%-2%** with a thyroglossal duct cyst may turn malignant! **Incidental Discovery**: **73.3% of these specific cancers** are discovered purely by chance during the final microscopic, post-operative analysis.

- Cancer in a thyroglossal duct cyst. TDGC

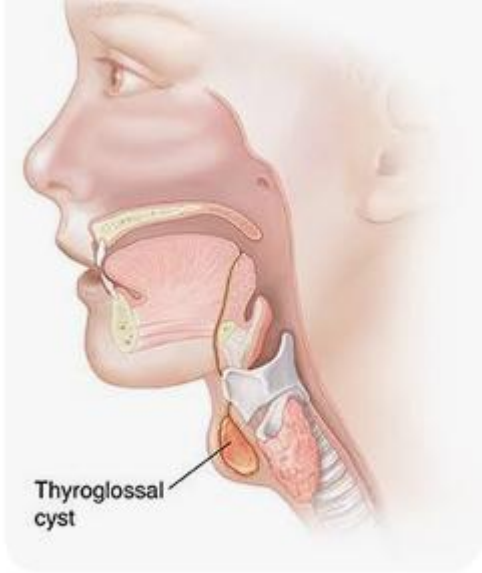
Adults: 30-40 years of age. Pediatric cancer <15%

Papillary carcinoma (>90%) –Very favorable prognosis!-

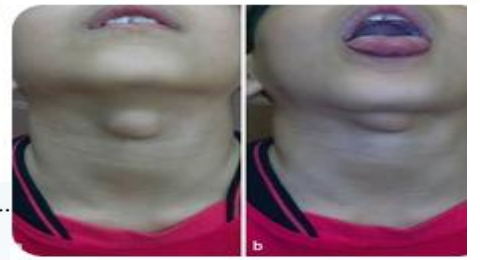
Squamous cell carcinoma (4.3%)

Follicular, Hurthle cell, Anaplastic, Mixed Neuroendocrine tumors.

"The thyroglossal duct cyst moves up and down when the person swallows or stick out the tongue"

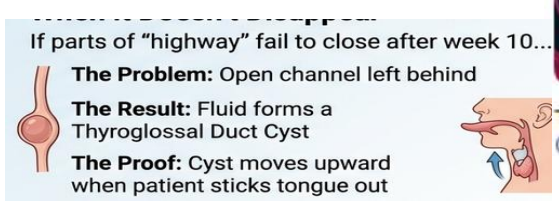


Text - Understanding Thyroglossal Cysts



Thyroglossal Cyst | Obgyn Key

Obgyn Key



00730 - Discriminator 1 - B-09.03.00.0004 L-1.0.0.1 A-09.03.02 S-3.2 TNM9LOV

Notes

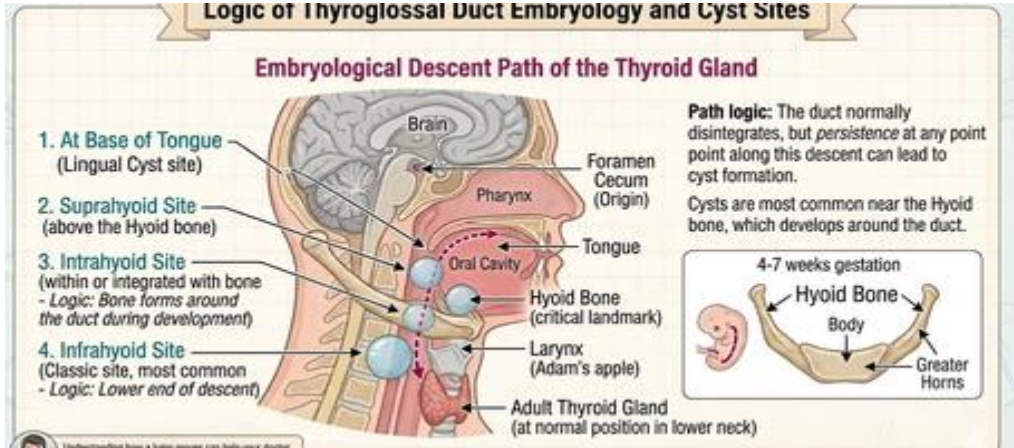
Note: **Schema Discriminator for C739**
* A schema discriminator is used to discriminate between thyroid gland and thyroglossal duct tumors with primary site code C739: Thyroid Gland. Code the site in which the tumor arose.

* **Thyroid gland (see code 1)**
Subsites include Thyroid, NOS
* **Thyroglossal duct (see code 2)**

Code	Description
1	Thyroid gland Thyroid, NOS
2	Thyroglossal duct cyst

C73 THYROID GLAND

C73.9 Thyroid gland
Thyroid, NOS
Thyroglossal duct



Thyroglossal duct cyst MALIGNANCY

Primary Surgical Management However, treatment is individualized as the stage of the malignancy can be combined with more extensive surgery, radiation and other options.

The Sistrunk Procedure

- **D**efinitive surgical standard of care.
- **Anatomical Scope**: During a Sistrunk surgery, surgeons cut and remove the **thyroglossal cyst**, the macroscopically visible tracking duct, a **central third of the hyoid bone**, and a **core of tissue** leading up to the base of the tongue.
  By resecting the midpoint of the bone ensures that tiny microscopic epithelial branches hidden near the bone cortex are entirely cleared.

Recurrence Rate:

By removing part of the hyoid bone drops the recurrence rate from 56% -70% to just 2.6% - 4.3%

Survival: **99.4% long-term survival rate** for papillary TGDC carcinomas.

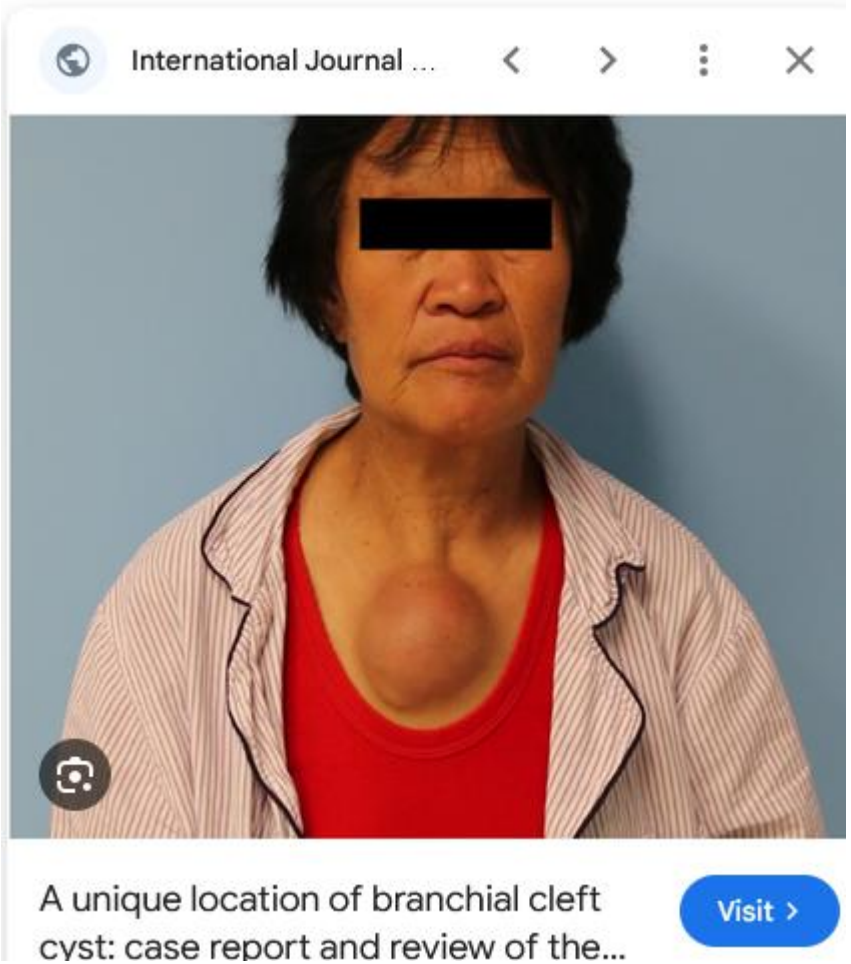
Branchial cleft and Meckel diverticulum as sites of neoplasms

Both "branchial cleft" and "Meckel diverticulum" are congenital abnormalities and as such are coded to categories Q18.0 and Q43.0 respectively in ICD-10. However, these anomalies create tissues in which neoplasms can arise. The codes C10.4, branchial cleft, and C17.3, Meckel diverticulum, are included in the topography section in ICD-O. The phrase "site of neoplasm" appears in parentheses after each term to indicate that they are to be used only when they are the site of origin of a neoplasm. ICD-O topography codes should not be used for these congenital anomalies unless a neoplasm arises in them.

C10.4 **Branchial cleft** *(site of neoplasm)*

C17.3 **Meckel diverticulum** *(site of neoplasm)*

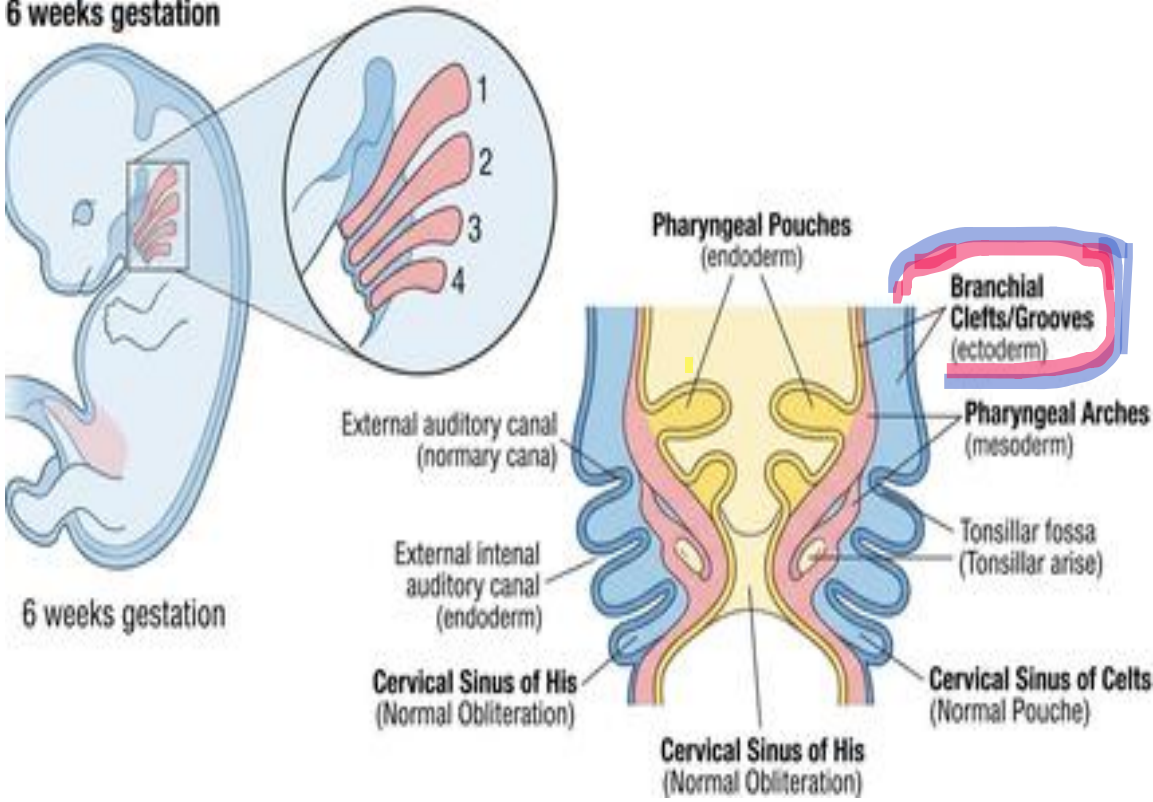
Branchial Cleft Cyst (Primary site)



C10.4 Branchial cleft *(site of neoplasm)*

Branchial (Pharyngeal)cleft or grooves

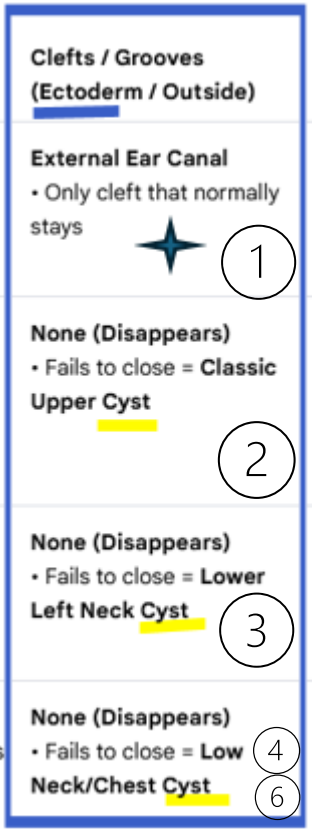
Number	Arches (Mesoderm / Muscles & Bones)	Clefts / Grooves (Ectoderm / Outside)	Pouches (Endoderm / Inside)
1	**Jaw & Ear Structures** • Mandible, Malleus, Incus • Muscles of mastication	**External Ear Canal** • Only cleft that normally stays	**Middle Ear & Eustachian Tube** • Connects the ear to the throat
2	**Upper Neck Structures** • Hyoid bone (body/lesser horn) • Muscles of facial expression	**None (Disappears)** • Fails to close = **Classic Upper Cyst**	**Tonsils** • Palatine tonsil lining
3	**Lower Neck Structures** • Hyoid bone (greater horn) • Stylopharyngeus muscle	**None (Disappears)** • Fails to close = **Lower Left Neck Cyst**	**Inferior Parathyroids & Thymus** • Glands migrate down to the chest
4 & 6	**Larynx / Voice Box** • Thyroid & Cricoid cartilages • Pharyngeal muscles	**None (Disappears)** • Fails to close = **Low Neck/Chest Cyst**	**Superior Parathyroids** • Glands settle on back of thyroid



Branchial Cleft Cysts develop on the SIDE of the neck, along the anterior border of the sternocleidomastoid muscle between the ear and collarbone, in close association with the carotid artery.

These embryonic PHARYNGEAL structures (Arches, Clefts and Pouches) act as the building blocks that grow into the face, neck, and throat.

Branchial cleft cyst CARCINOMA…



- Rule of Exclusion: Doctors must aggressively rule out an occult head, neck, thyroid primary or other primary possibility.

Strict Diagnostic Criteria (Martin's Criteria): To be called a primary branchial carcinoma, the tumor must be located anatomically in a branchial tract, must show carcinoma arising directly out of benign cyst lining, and **the clinician must rule out any other head/neck primary for at least 3-5 years.**

Many of suspected cases actually represent cystic lymph node metastasis from an occult primary cancer.

- We have received some valid Branchial cleft cyst cancers reporting to FCDS

Abstract

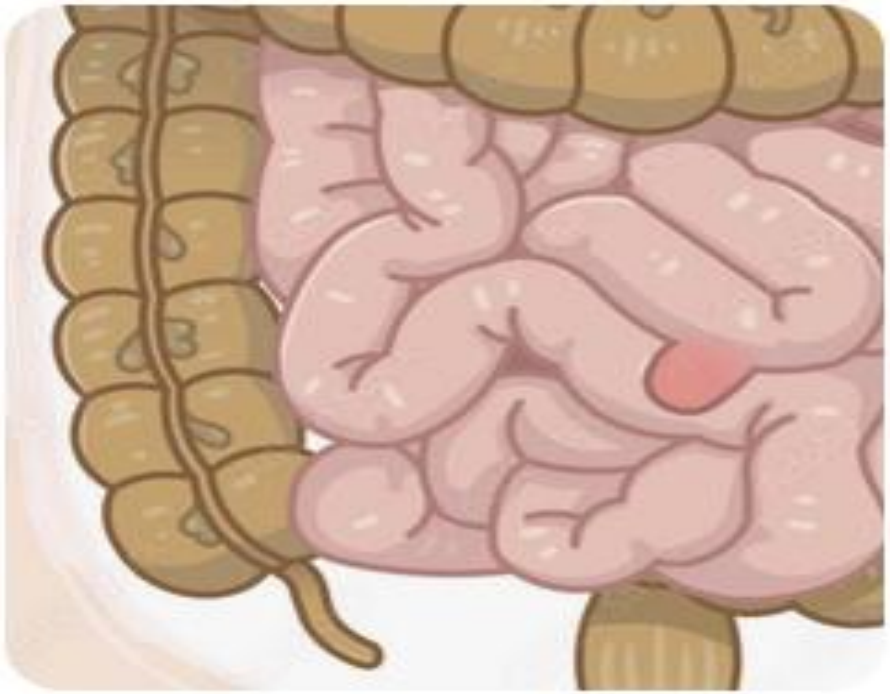
C10.4 Branchial cleft *(site of neoplasm)*

Branchial cleft cyst carcinoma (BCCC) is an extremely rare malignancy originating from cells within the branchial cleft cyst wall. A 73-year-old man presented with a cystic mass with cellulitis mimicking abscess initially and recurred 3 years later as complex cystic lesion in right neck level II with multiple necrotic ipsilateral lymphadenopathy. The pathological diagnosis of cystic lesion was squamous cell carcinoma, suggesting possibility of originating from branchial cleft cyst. There was no identifiable primary cancer elsewhere by a thorough evaluation; eventually final diagnosis was branchial cleft cyst carcinoma to meet the modified criteria of Khafif et al. Up to the present time, there has been no evidence of recurrence. Although the BCCC is very rare, accurate diagnosis is important to plan proper treatment for patient. This report should help increase awareness of BCCC, which should be included in the differential diagnosis of a cystic neck mass.

https://brieflands.com/journals/ijradiology/articles/18127

Meckel's Diverticulum (Primary site)

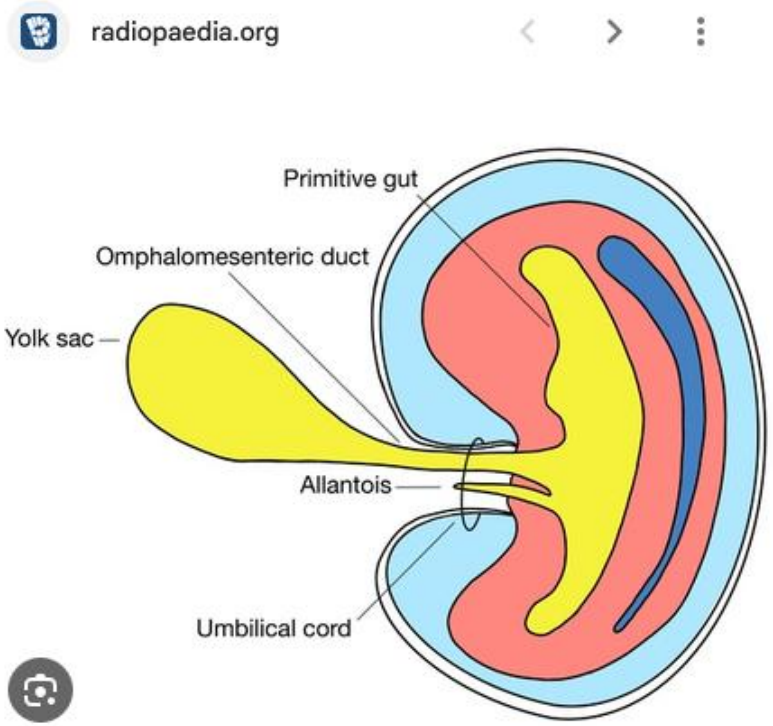
C17.3 **Meckel diverticulum** *(site of neoplasm)*

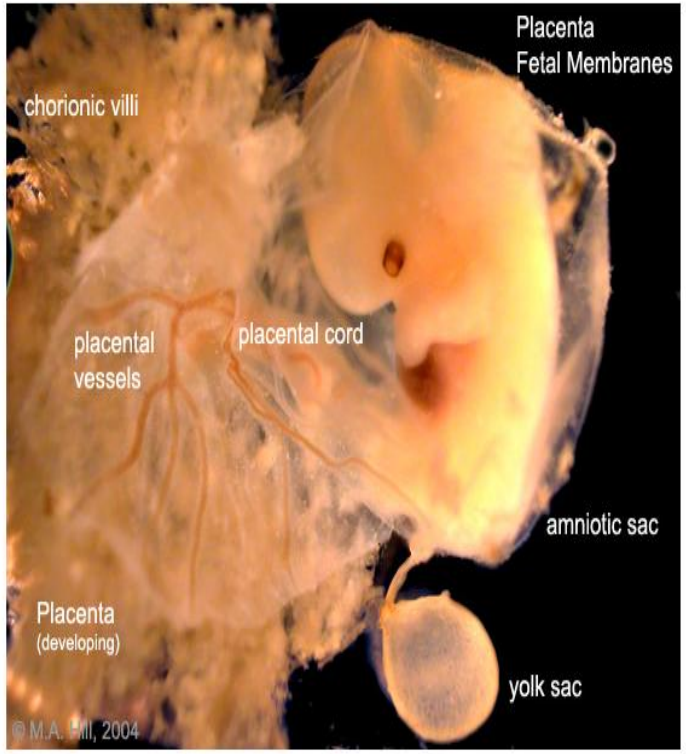


Meckel's Diverticulum -...

TeachMe Surgery

A **diverticulum** is a small, bulging pouch or sac





Omphalomesenteric duct or vitelline duct is a temporary tube in an embryo that connects the yolk sac to the developing gut (**nutrient** transfer and guide to **rotation** of human digestive tract).
It should close and disappear by 6-9 week of gestation. If it does not go away completely, it can cause a Meckel diverticulum

Meckel's Diverticulum

Embryonic Origin: Results from the incomplete closure of the omphalomesenteric (vitelline) duct.

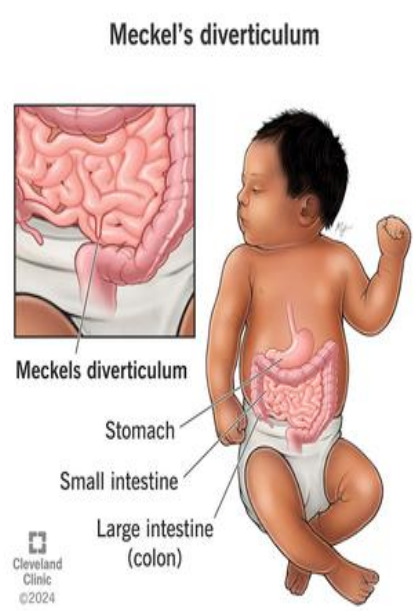
- **The "Rule of 2s"**
- Found in **2%** of people
- About **2 inches** long
- Located within **2 feet** of the large intestine connection (ileocecal valve)
- Most often causes problems before **2 years** of age
- Males **2 times** more than females

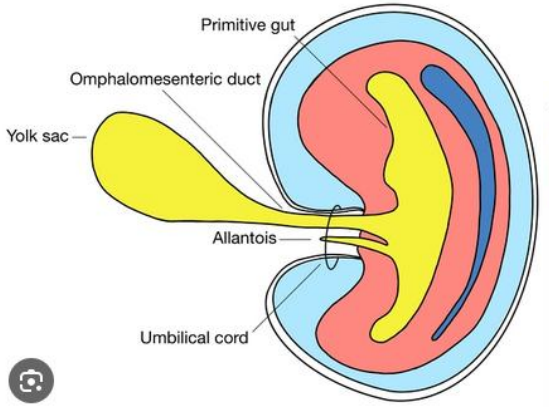
Symptoms

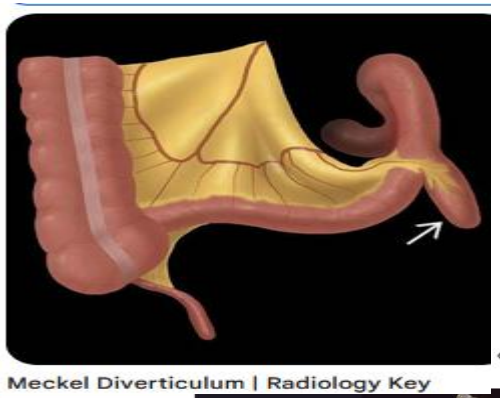
- **Painless bleeding** from the rectum (most common in children)
- **Stomach pain** and tenderness if the pouch cause the bowel to twist causing obstruction.
- **Blockage** in the gut causing vomiting or bloating
- **Diverticulitis** the pouch can become inflamed and infected

Treatment: Surgery **remove the pouch** if it causes bleeding, pain, or blockages.

A Meckel diverticulum can contain misplaced stomach tissue that makes **hydrochloric acid**, , which harms the nearby intestine and causes bleeding or painful ulcers.






Meckel Diverticulum | Radiology Key

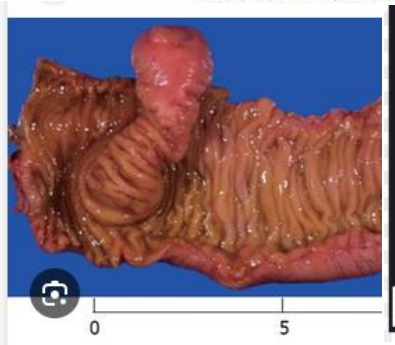
Malignancy Risk: Over 95% are benign

Cancer develops in only **0.5% to 3.2%**

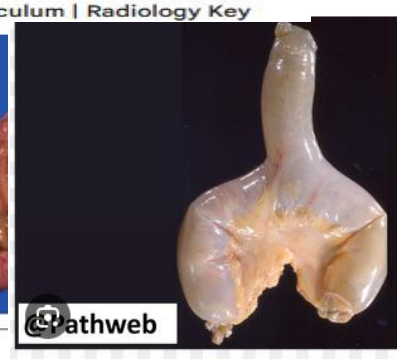
Neuroendocrine, Gastrointestinal stromal tumor (GIST), Adenocarcinoma

15.8% Metastases from secondary location

Age: **60s (median age)**



Inverted Meckel's diverticulum



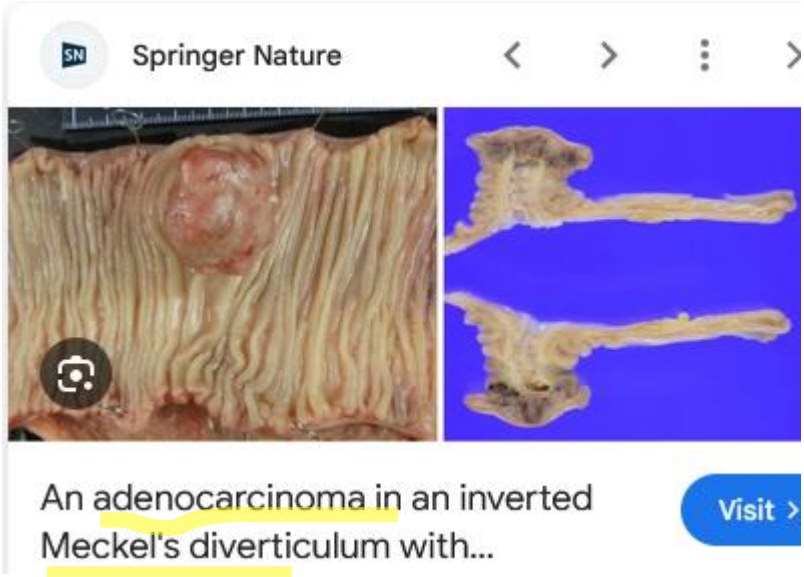
Intestine – Meckel's Diverticulum –

Meckel’s Diverticulum Cancer

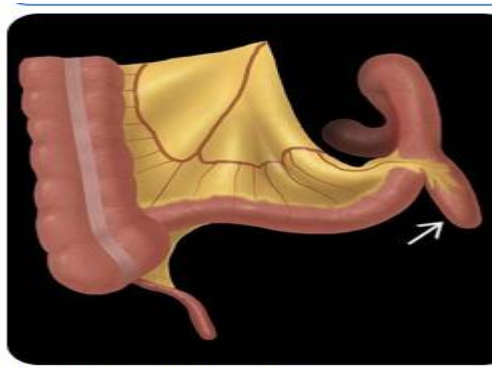
C17.3	Diverticulum, Meckel *(site of neoplasm)*

Meckel's diverticulum is a small birth defect pouch in the wall of the lower small intestine.
Incidence: Occurs in only **0.5% to 3.2%** of all Meckel's diverticulum cases
Tumor Types:

- **Neuroendocrine/Carcinoid tumors:** 33% to 44%
- **Sarcomas / GISTs** (Gastrointestinal Stromal Tumors): 18% to 35%.
- **Adenocarcinomas:** 6% to 16%
- Rare: **lymphomas** or **squamous cell carcinomas**.

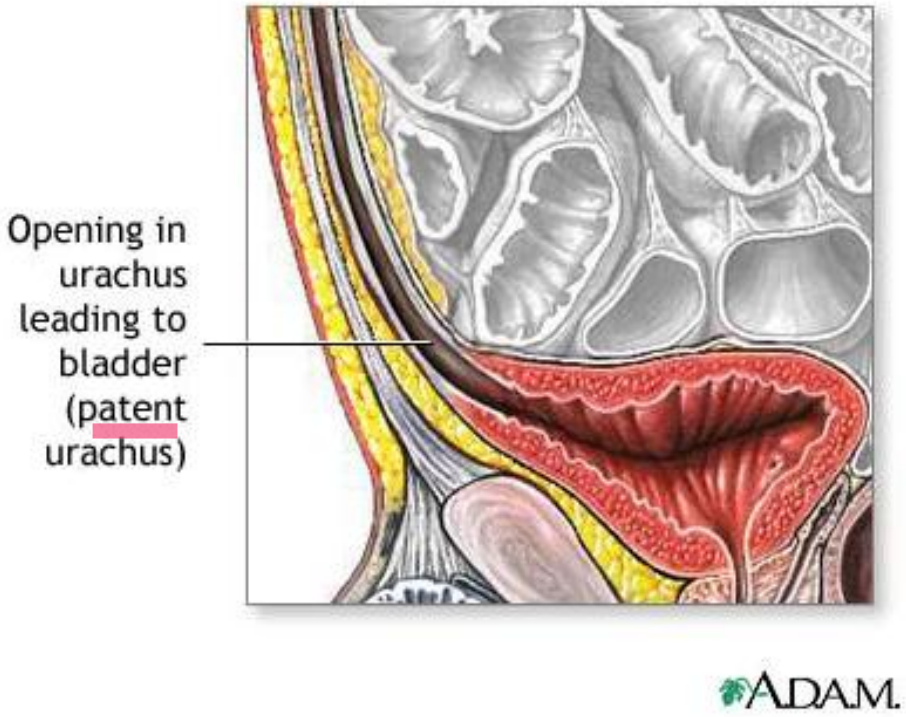


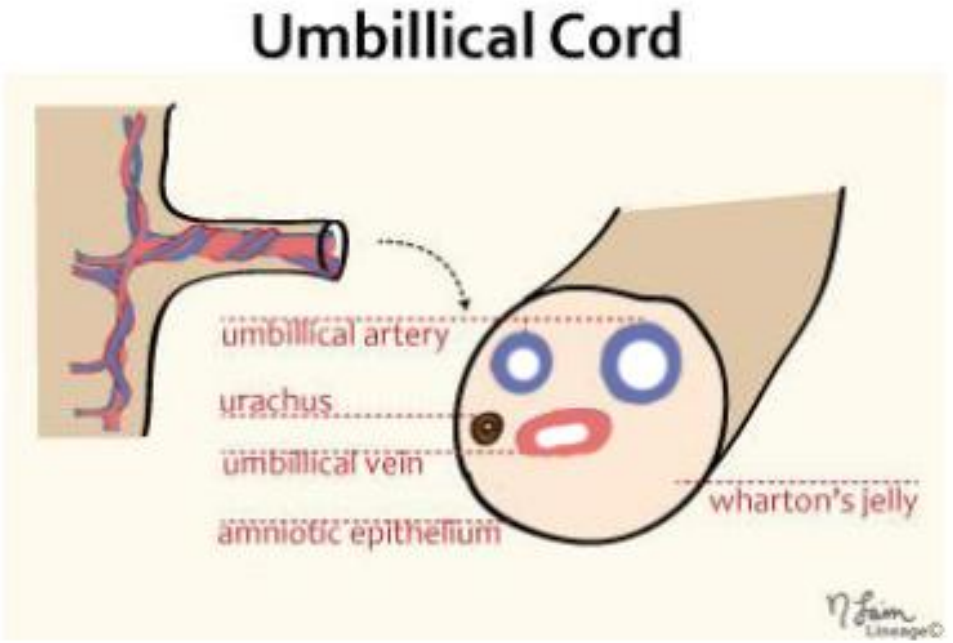
- **Clinical Presentation & Symptoms**
- **Asymptomatic**: Most tumors are completely silent until advanced stages.
- **Constitutional**: Unexplained weight loss and chronic fatigue.
- **Mimicry**: Frequently misdiagnosed as acute appendicitis due to localized RLQ pain.
- **Gastrointestinal Bleeding**: Melena or bright red blood per rectum.
- **Bowel Obstruction**: Caused by tumor mass, intussusception, or volvulus.


Meckel Diverticulum | Radiology Key

Urachus (Primary site)

C67.7	Urachus

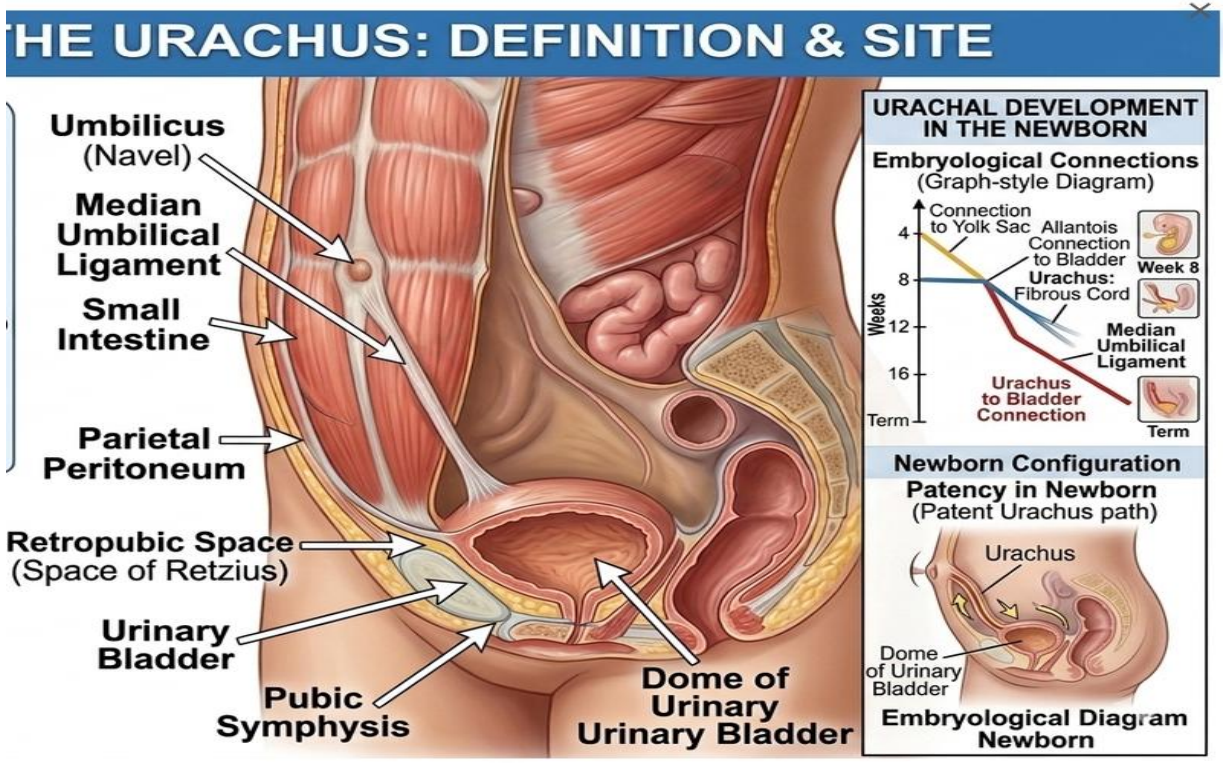




URACHUS

Adult Structure: Also known as the **Median Umbilical Ligament** (anatomical site/name).
Function (Embryo): Allowed drainage of fetal urine.
Function (Adult): None; vestigial structure.

The urachus is the internal remnant of the embryonic **Allantois**, forming a tube.



- **In the Fetus (as the Urachus):** Drains fetal urine from the dome of the developing bladder into the **allantois** and out through the umbilical cord.
  Once it goes through the placenta to the mother's bloodstream, the mother's kidneys filter this waste.
- **In the Newborn (or before birth):** Undergoes rapid closure (**obliteration**) after birth to prevent urine from leaking out of the belly button.
- **In the Adult:** Persists strictly as a vestigial, fibrous remnant with **no active physiological function**, though it serves as an anatomical landmark on the anterior abdominal wall.

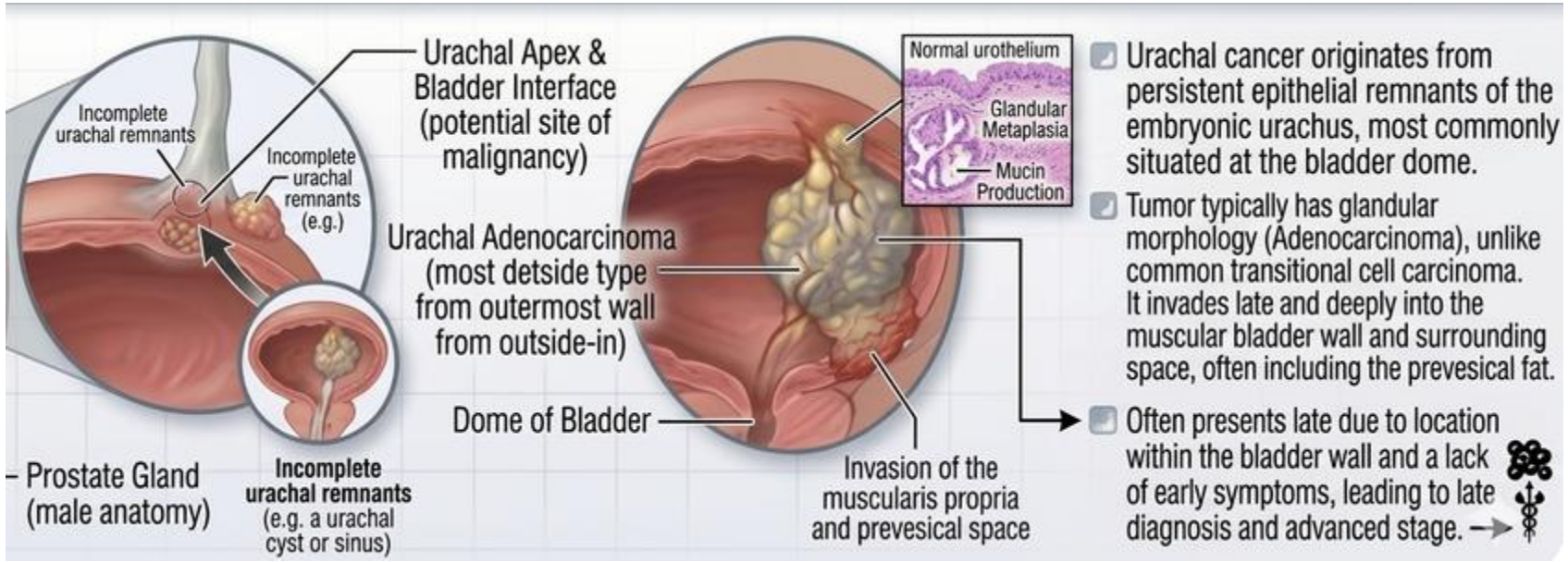
C67.7 **Urachus**

C67.8 **Overlapping lesion of bladder**
(see section 4.2.6)

C67.9 **Bladder, NOS**
Bladder wall, NOS
Urinary bladder, NOS

The urachus turns into a fibrous cord of tissue that extends from the dome of the urinary bladder up to the umbilicus (belly button). In an adult body, it is a non-functional remnant of embryonic development known anatomically as the **median umbilical ligament**.

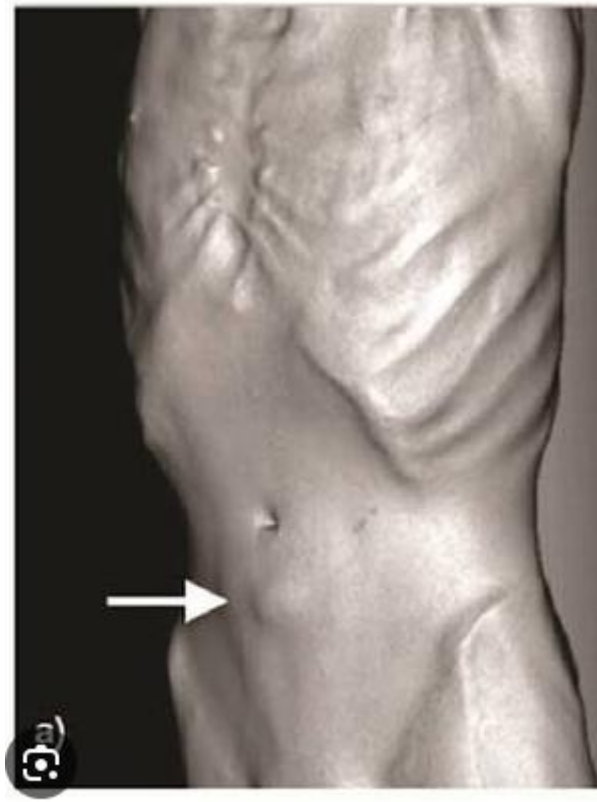
Urachal Cancer



Types of Urachal Cancer:
-Adenocarcinoma: 85- 90% of cases.
Subtypes include mucinous, enteric, and signet-ring cell.
-Urothelial, Squamous cell carcinoma, Neuroendocrine carcinoma, and sarcoma.

Incidence 1 in 5 million people. The median age at diagnosis: **50 and 60 years old.**

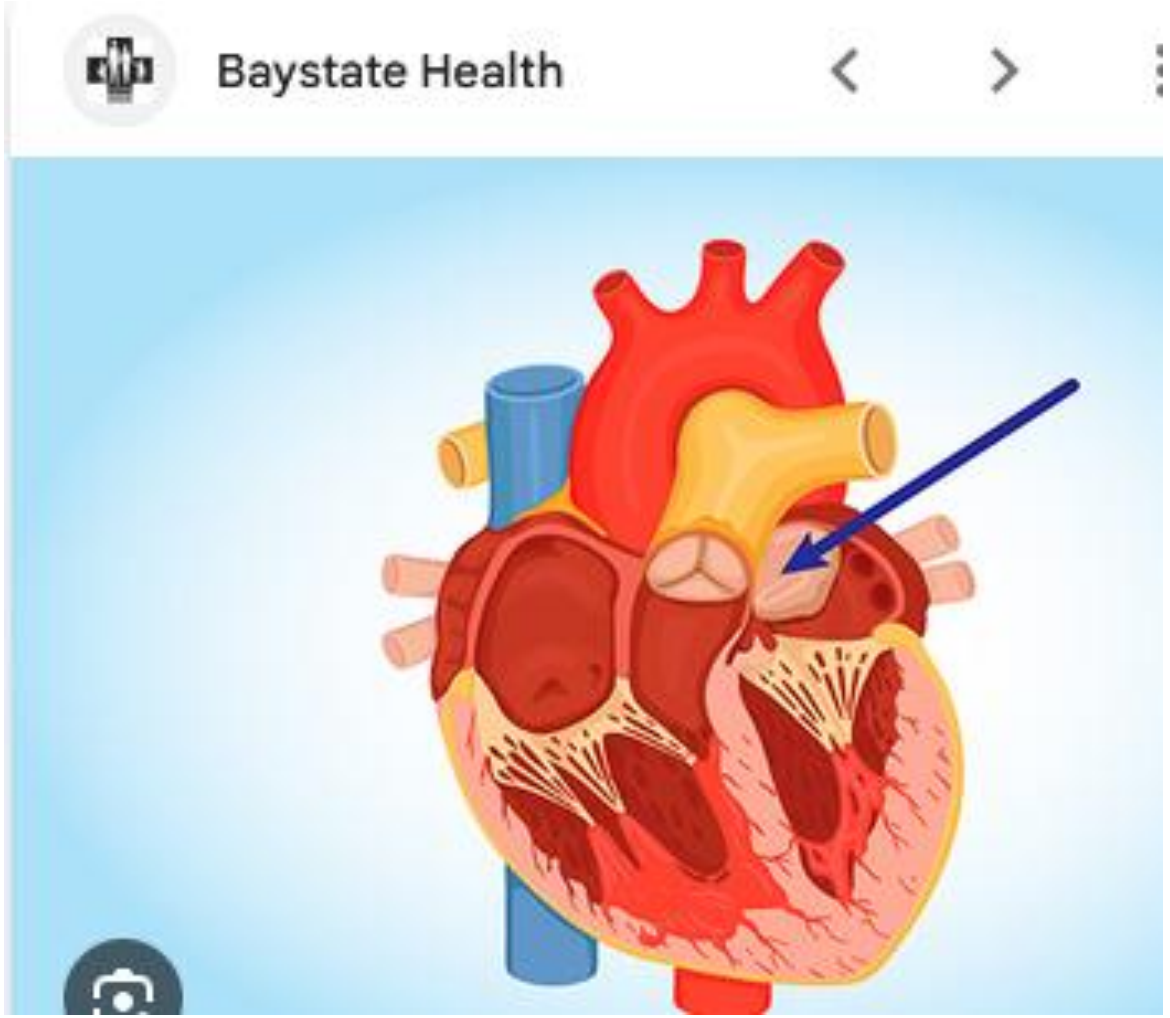
Wiley Online Library

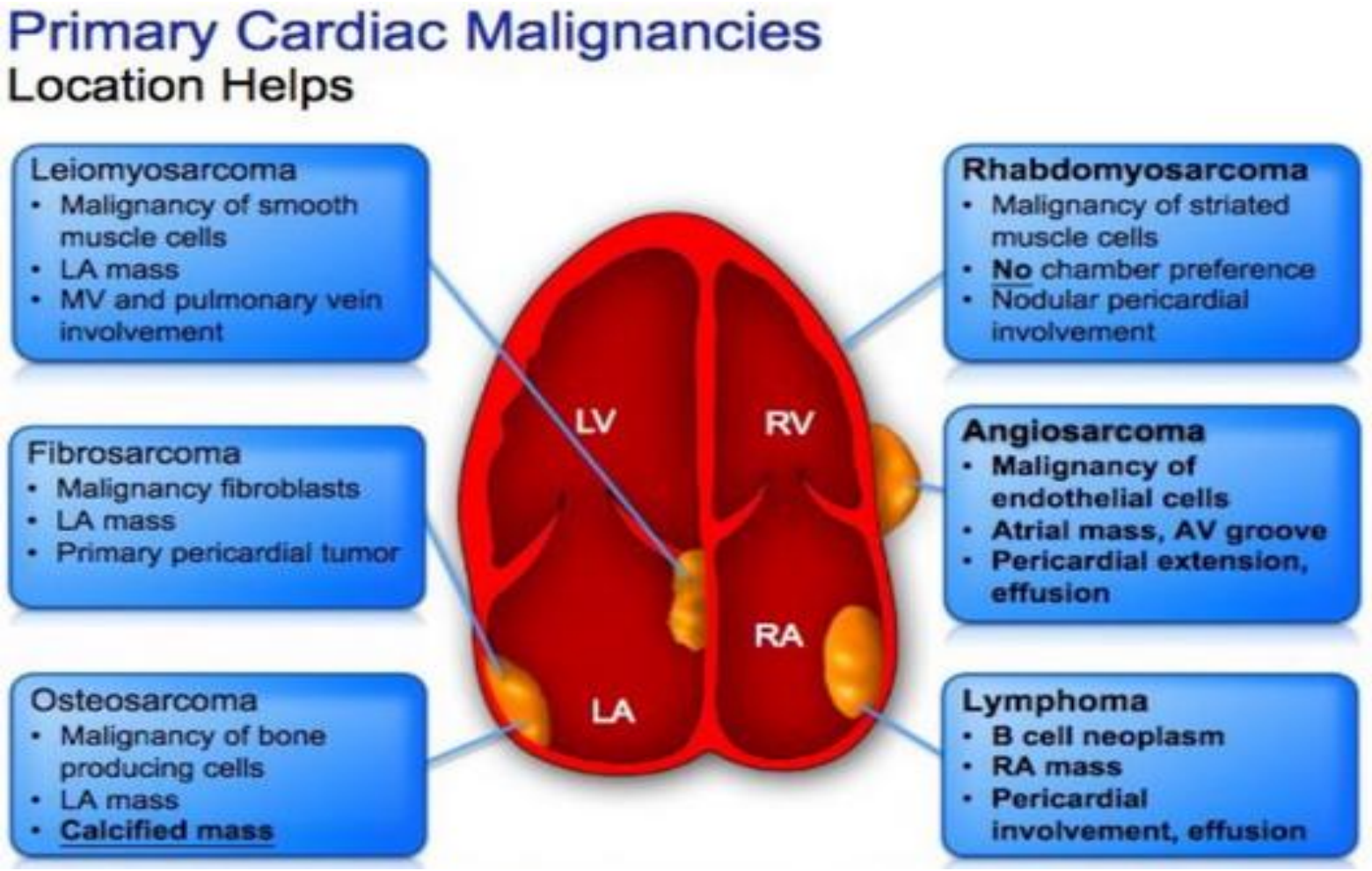


Urachal Adenocarcinoma: A Case

We present a case of a 13-year-old female who experienced hematuria, dysuria, and abdominal pain persisting for over 4months. A CT scan revealed a mass extending from the bladder wall, involving an adjacent bowel loop, and associated with intra-abdominal lymphadenopathy. Debulking surgery was performed, and a histopathological examination confirmed the diagnosis of urachal adenocarcinoma.

Heart (Primary site)





Heart primary site

- Most primary Heart tumors are **Benign**: 75% -90%

  -Cardiac Myxoma, Cardiac Rhabdomyoma, Papillary Fibroelastoma.

  They can be life-threatening or fatal if they are left untreated: Progressive severe heart failure, arrythmias, strokes, embolisms, blood flow obstruction.

- Primary Cardiac **Malignancies**: 10% to 25%

  -Cardiac **SARCOMAS**: 95%

  AngioSARCOMA (30%-43%)
  LeiomyoSARCOMA (6%-9%)
  RhabdomyoSARCOMA (3%–9%)
  OsteoSARCOMA (1%–4%)
  FibroSARCOMA (< 3%), and -----

  -Primary **LYMPHOMA**: 1%–5%

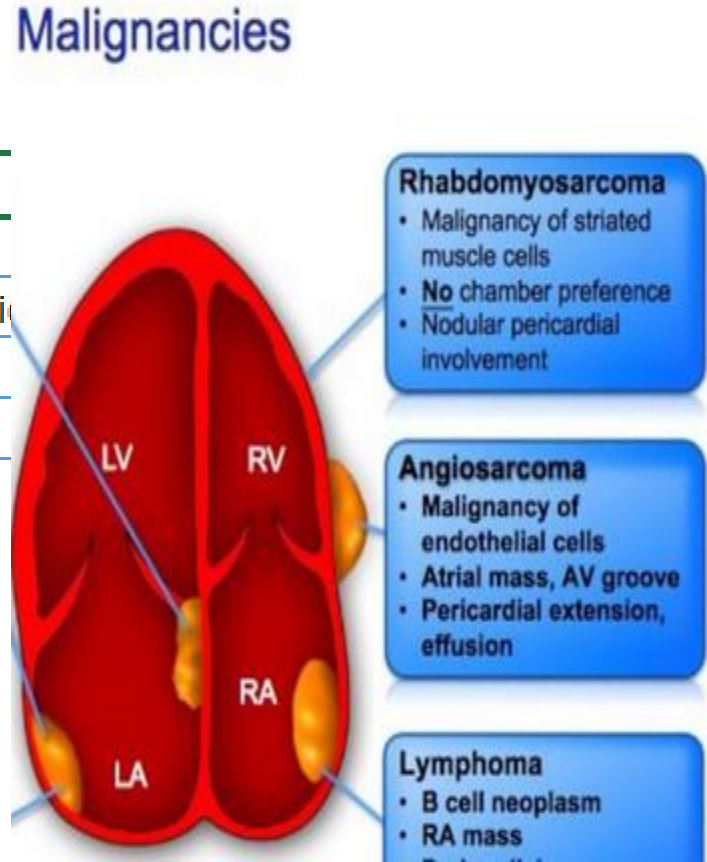
CARDIAC Embryonal Rhabdomyosarcoma (ERMS)

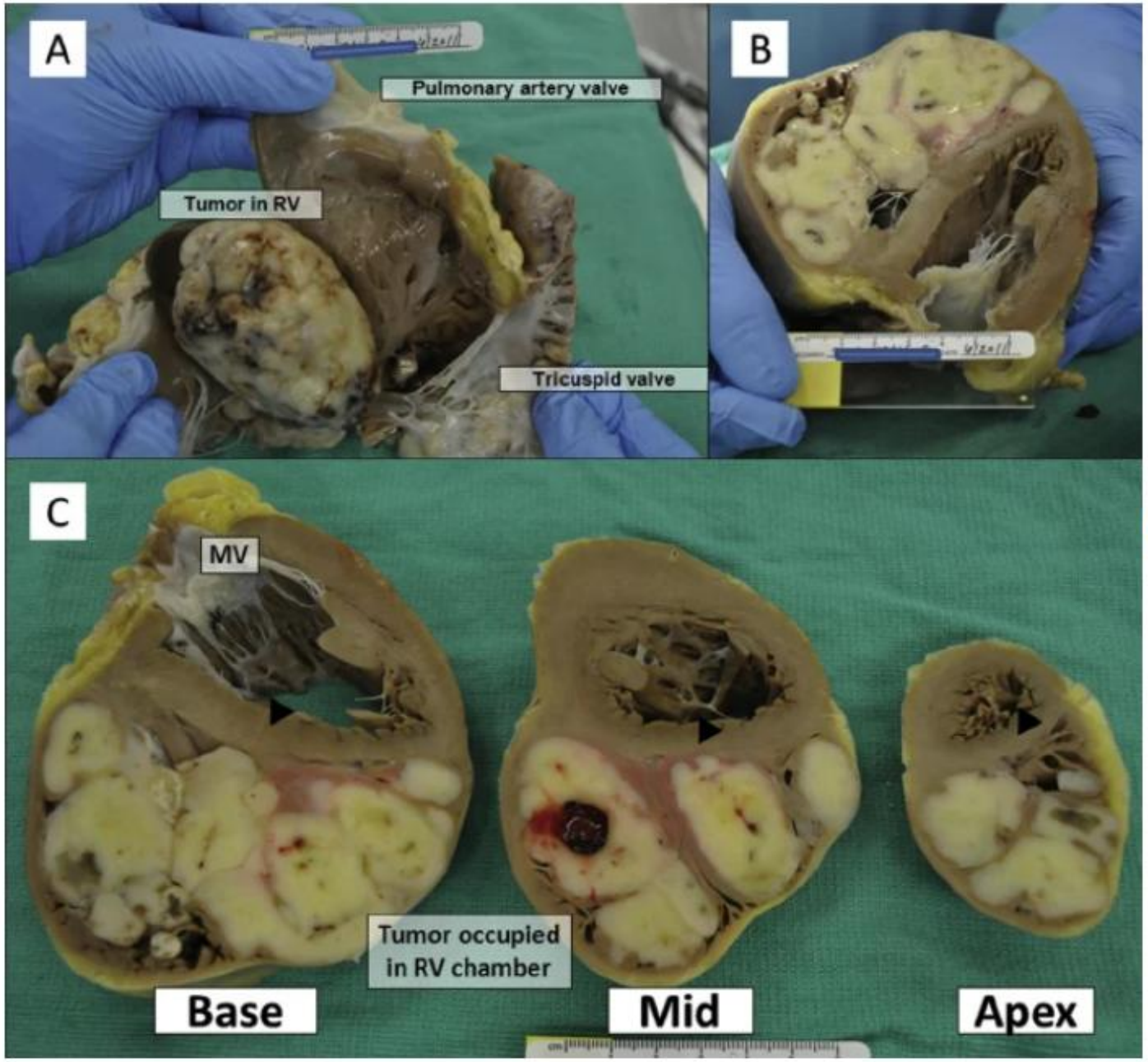
Text - Dx Procedures - Pathology Report

12/11/21 BIOPSY OF RIGHT PARAPHARYNGEAL SPACE MASS: NO EVIDENCE OF MALIGNANCY.1/19/22 INTRACARDIAC MASS RESECTION: RHABDOMYOSARCOMA, EMBRYONAL W/ANAPLASTIC FEATURES, FOXO1 NEGATIVE.

8910/3	8910	3	Preferred	Embryonal rhabdomyosarcoma, NOS
8910/3	8910	3	Synonym	Rhabdomyosarcoma, embryonal type
8910/3	8910	3	Related	Embryonal rhabdomyosarcoma, pleomorphi
8910/3	8910	3	Related	Sarcoma botryoides
8910/3	8910	3	Synonym	Botryoid sarcoma

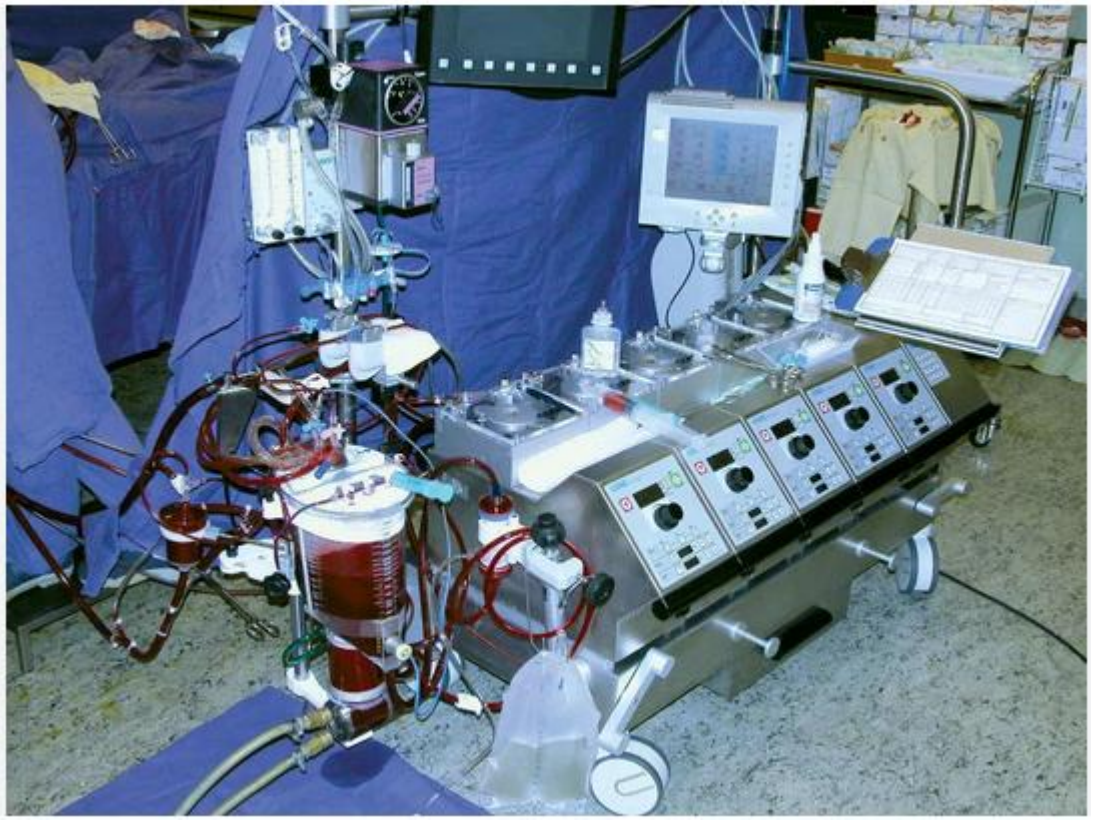
- Usually affects infants and young children, but it can affect adults.
- They usually affect the VENTRICULAR walls, but in adults it may arise from the ATRIAL cardiac walls.
- It metastasizes. Long term prognosis is poor. Highly lethal!



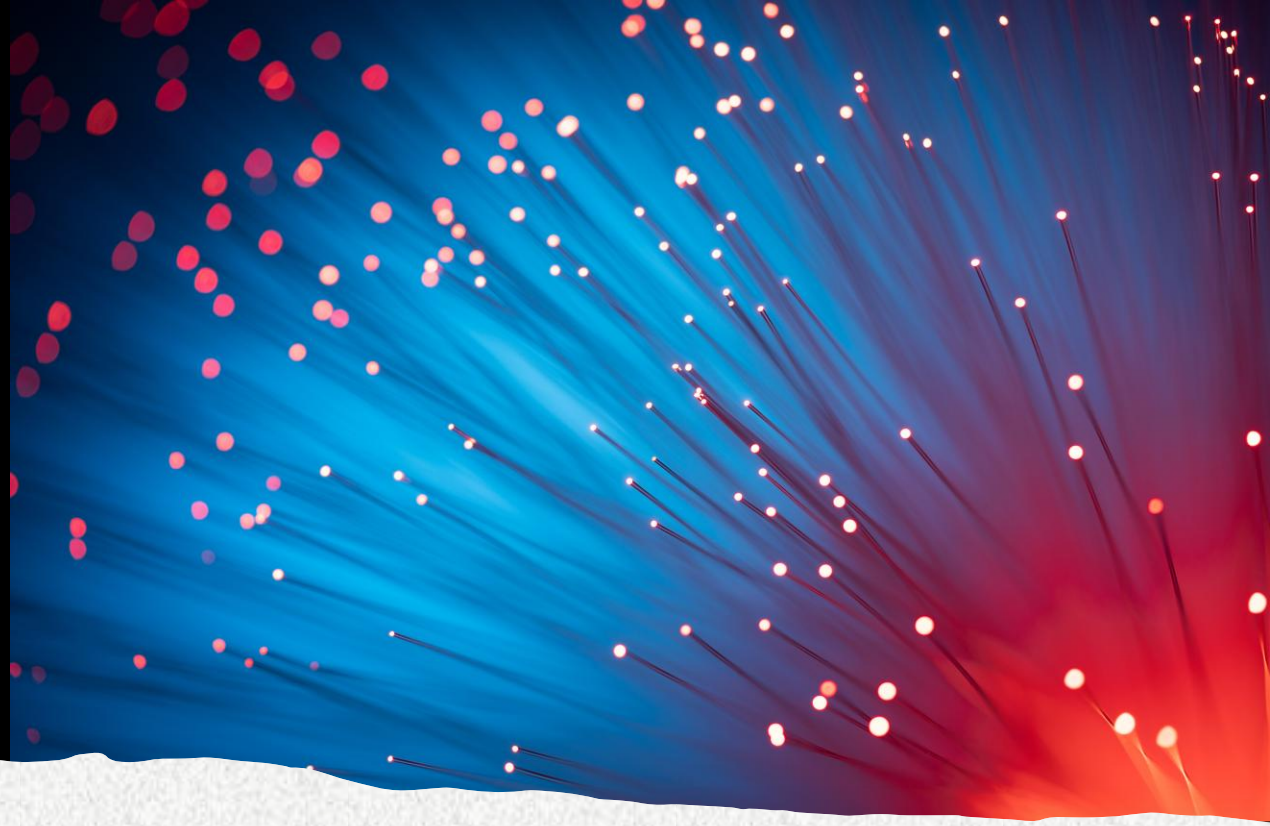


Gross examination of resected heart. (A) Right ventricle tumor. (B) Right ventricle cross section. (C) Successive cross sections demonstrating near obliteration of right ventricular chamber. (MV ¼ mitral valve; RV ¼ right ventricle.)

Heart-lung machine



Heart-Lung Machine is a cardiopulmonar bypass machine which temporary takes over the functions of the heart and lungs while surgeons perform open-heart surgery. It adds oxygen, removes carbon dioxide, and pumps blood to the rest of the body while working on the heart.



Thank you for your time!

- **Embryology Trajectory**: Foramen cecum migration and tract anatomy details validated via the NCBI StatPearls Bookshelf Head and Neck Guide.
- **Anatomical Reference Data**: Pathological markers and imaging criteria cross-checked via the Radiopaedia Reference Library.
- **Clinical Presentation Models**: Patient presentation features and adult mass development profiles reviewed through the Cleveland Clinic Medical Education Library.
- **Tumor Morphology Distributions**: The 92.1% papillary and 4.3% squamous cell subtype statistics are reported by Pathology Outlines Core Database.
- **Incidental Identification Incidence**: The 73.3% incidental detection metric is pulled from surgical pathology reviews inside the National Institutes of Health PMC Archive.
- **Surgical Outcomes & Failure Percentages**: The 55.6% simple excision failure rate versus the ~5% Sistrunk success rate is cataloged in the PubMed Head & Neck Journal Studies.
- **Survival and Recurrence Calculations**: The 99.4% survival rate and 4.3% disease recurrence metrics are confirmed via the StatPearls Surgical Oncology Protocols.

- Medscape
- https://www.sciencedirect.com/science/article/pii/S221026122500032X
- https://www.journalmc.org/index.php/JMC/article/view/1033/677
- https://pmc.ncbi.nlm.nih.gov/articles/PMC12796787/
- https://my.clevelandclinic.org/health/diseases/22351-thyroglossal-duct-cysts
- https://www.vumc.org/cbmes/publication/papillary-type-carcinoma-thyroglossal-duct-cyst-case-conservative-management
- https://pmc.ncbi.nlm.nih.gov/articles/PMC7201451/
- Merck Manual
- Medscape
- https://pmc.ncbi.nlm.nih.gov/articles/PMC6068795/
- https://pmc.ncbi.nlm.nih.gov/articles/PMC7547967/