The spectrum of sonographic appearances of normal and abnormal pediatric cervical thymus: what we need to know

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Authors: M. Vakaki\textsuperscript{1}, K. Kalogerakou\textsuperscript{1}, G. Karapostolakis\textsuperscript{1}, D. Grigoraki\textsuperscript{1}, K. Boursiani\textsuperscript{1}, C. Koumanidou\textsuperscript{2}\textsuperscript{;}; \textsuperscript{1}Athens/GR, \textsuperscript{2}P.Faliro-Athens/GR
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Learning objectives

- To familiarize pediatric radiologists with the normal embryology of thymus and the presence of thymic remnants along the path of its descent.

- To illustrate a wide range of congenital thymic conditions, and to emphasize their key sonographic findings.
Background

Palpation of a neck mass, a common disorder in infants and children, is usually alarming, leading to increased anxiety for families.

On the other hand, incidental sonographic detection of a thyroid abnormality in a child undergoing neck sonographic examination for non-thyroid indications, may pose a clinical dilemma regarding management.

Pediatric radiologists play a major role in differentiating:
- normal variants, thus avoiding unnecessary follow-up and investigations, and
- pathologic conditions, needing further investigation.

Given the advances in US technology, Ultrasound is the initial and usually the only imaging modality of choice.

It does not require sedation such as with MR and CT,
it does not involve ionizing radiation such as with CT, and
it does not utilize contrast media such as with MR and CT.
The word *thymus* derives from the Greek *thymos* meaning 'soul' or 'spirit'. Galen theorized that the function of the thymus was that of a cushion, to protect the mediastinal vessels from the overlying sternum. It was in the late 1700s that an association was suggested between the development of the lymphatic system and the thymus. It is currently accepted that the thymus serves as a central lymphoid organ that is responsible for the development and maturation of T-and B-lymphocytes and induction of self-tolerance.

**Embryology and anatomy of the thymus gland**

The knowledge of thymic embryology is essential to understand how:

- the sonographic demonstration of cervical thymic tissue corresponds to its abnormal caudal migration, and

- the various thymopharyngeal duct remnants sonographically appear.

**Thymus-Embryology:**

The 3d and 4th pharyngeal pouches in the embryo are characterized by the so-called dorsal and ventral wings. The thymus develops mainly from the ventral wing of the 3d pharyngeal pouch on each side. The dorsal wings of the same pouch give rise to the inferior parathyroid glands. The thymic primordia that develop from the 4th pharyngeal pouch, even at the time of its formation, are likely to be quite small and rudimentary. They may give rise only to vestigial tissue masses. The superior parathyroid glands develop from the dorsal wings of the same pouches.

The primordia thymus appears in the beginning of the 6th week of fetal life. By the end of this week the connections of the primordia with the pharyngeal pouches are severed. Each primordium elongates caudally and medially as a tubular structure (*thymopharyngeal duct*). By the 8th week, the bilateral thymic primordia fuse in the midline and start to slide down under the sternum in the superior mediastinum, where they lie in contact with the parietal pericardium. The path of descent involves migration deep to the thyroid gland and sternocleidomastoid muscle along the carotid sheath. In normal development, the inferior portions of the thymopharyngeal stalks enlarge, while the proximal portions form epithelial cords that eventually atrophy. The final location of the thymus is usually entirely in the anterior mediastinum, between the sternum, parietal pericardium and thoracic inlet. The thymus is attached to the thyroid by the thyrothymic ligament.

**Thymus-Anatomy:**
Each of the two lobes of the thymus is divided by primary connective tissue septa carrying blood vessels to parenchymal lobules, which are composed of a cortex and medulla, both of which are developed by the 12th week. The lobules are further divided by secondary connective tissue septa carrying blood vessels from the surface of the cortex to the corticomedullary boundary. The cortex contains only capillaries, whereas, the medulla has both small arteries and arterioles. The thymus increases in size until puberty, where it can range from 30 to 40 grams. After puberty, there is gradual fatty involution that replaces the lymphoid components.

Pathogenesis of the cervical thymus (Fig. 1)

I) The mechanisms that account for a retained solid cervical thymus include:

- IA) sequestration of thymic tissue during descent,
- IB) failure of involution, or
- IC) arrest in migration.

IA) In the case of sequestration, thymic tissue becomes implanted in the neck from the main gland during its normal fetal descent and form a solitary mass which in essence is a true aberrant thymus in the neck. Such nests of aberrant thymic tissue may be found anywhere along the expected path of descent from the angle of the mandible to the mediastinum.

Aberrant thymus is commonly located in the lateral neck or in the suprasternal region, and is usually an asymptomatic, incidental finding.

Ectopic thymic tissue can be found in any other location, such as the pharynx, trachea, posterior neck or mediastinum, and esophagus. Ectopic thymus can occasionally be life-threatening, such as with airway obstruction.

The paths of descent of the thymus and thyroid are closely related due to the proximity of the thyroid diverticulum and the 3d branchial pouches. Therefore, thymic tissue can get sequestered within the thyroid gland, resulting in ectopic intrathyroid thymus. Until recently, intrathyroid thymus was considered rare and a small number of cases was reported in the literature.

Note: The terms "aberrant" and "ectopic" thymic tissue have been used interchangeably, referring to any thymic tissue found in a location other than the anterior mediastinum.

IB) Sometimes after descent, the upper ends of the gland, instead of involuting, form separate accessory lobes or persistent cords.
IC) Rarely, the **thymus fails to descend from the neck or descends incompletely**. Naturally, it then remains wholly or partially in the cervical region. This situation is almost always asymptomatic and usually incidentally discovered.

- Another presentation of cervical thymus is **hyperplasia of the normally situated thymus gland secondary to infections or vaccinations**.

There are no data to indicate that the incidence of malignant transformation of cervical thymus is any greater than in normal-positioned mediastinal thymus.

II) Cystic thymic remnants

Also, as the thymopharyngeal duct undergoes atrophy, when the duct fails to involute, either a:

IIA- **thymic cyst**, or

IIB- thymopharyngeal duct cyst (**persistent thymopharyngeal duct**).

may occur anywhere along its course from the angle of the mandible to the upper mediastinum.

Given that the parathyroid glands arise from the 3d and 4th pharyngeal pouches, too, parathyroid tissue may appear anywhere near or within the thymic remnants.

Up to 50% of cervical thymic cysts will be continuous with the mediastinal thymus. Cervical thymic cysts are intimately associated with the carotid sheath; they frequently splay the carotid artery and jugular vein, particularly when they involve the suprahyoid neck.

Important: As with branchial apparatus anomalies, the location is the key to suggesting the diagnosis. In a child, any cystic mass that involves the neck and also extends to the anterior mediastinum should certainly suggest a cervical thymic cyst.

Cervical thymic cysts are clinically important, as they have a similar appearance to other congenital neck masses and may demonstrate progressive growth. The underlying pathogenic mechanisms in the development of cervical thymic cysts remain controversial. In 1938, Speer proposed 5 theories. They may represent: 1. remnants of branchial clefts or the thymopharyngeal tract, 2. neoplastic change in the lymphoid or surrounding tissues, 3. sequestration of thymic tissue during migration, 4. cystic degeneration of Hassall's corpuscles, 5. they may arise from lymphoid tissue that has arrested in various
stages of thymic development. The two favored explanations are the persistence of thymopharyngeal tracts (congenital) and degeneration of Hassall's corpuscles within ectopic thymic remnants (acquired).

A classification system proposed for cystic thymic remnants differentiates these lesions based on structures of origin. **True cysts** are those lesions that originate in maldevelopments of the thymopharyngeal duct and may span the entire length of the neck with or without a patent fistula tract to the pyriform sinus. When these masses span the length of the neck from mandibular angle to clavicle, they are termed thymopharyngeal duct cysts. **Mixed cysts** contain elements of second brachial cleft and third pharyngeal pouch origin and are considered thymic fistulas. Those lesions derived from brachial cleft remnants, with incidental thymic tissue present, are referred to as **false thymic cysts**.

Congenital thymic cysts compose <1% of pediatric cervicothoracic masses and are essentially benign. 80-90% of patients with cervical thymic cyst are asymptomatic and have only a painless swelling. Respiratory symptoms such as dyspnea, hoarseness, stridor, and dysphagia are reported in 6-13% of patients. Progressive airway obstruction secondary to a rapidly enlarging cervical thymic cyst has been reported in neonates. Malignant transformation has been reported in adults but not in children, possibly due to the fact that a thymic cyst contains no active solid thymic tissue. There has been no reported recurrence in a child, after surgical excision.

**Sonographic findings**

Sonography represents the imaging method of choice for the study of the thymus in early childhood. Further investigation (imaging and/or FNA) is rarely indicated.

**Thymus (normal thymus)**

The sonographic appearance of the thymus depends on the age of the child: its echogenicity and coarseness increase with advancing age (a fine mixture of remaining lymphoid tissue and fat).

Knowledge of the sonographic patterns of the aging thymus enables a diagnosis of cervical thymic rests.

Thymus has a characteristic and unique appearance: branching or linear echogenic septa and multiple discrete echogenic foci throughout the gland, believed to represent connective tissue septa and their associated vasculature (starry sky appearance). The cortex of the lobules is typically hypoechoic versus the more echogenic medulla. The unique ultrasound pattern allows for easy differentiation of thymus from liver, spleen, and thyroid. In cases of thymic tissue near the mandible, one can also differentiate
thymus from salivary glands (including parotid and submandibular) that are typically more homogeneous with fine internal echoes.

*Important:* The thymus is very pliable. It does not compress or displace the adjacent anatomic structures. The same characterizes the cervical thymus.

I) **Solid Cervical thymus**

**Aberrant cervical thymus** and **Ectopic thymic tissue (Fig.2)**

Ultrasound demonstrates normal thymus in its expected location in the anterior mediastinum, as well as, the aberrant or ectopic thymus in the neck.

*Important:* the aberrant, ectopic and native thymus demonstrate the same sonographic characteristics. Ultrasound can be used to confidently identify tissue as being of thymic origin.

Unique linear echogenic septa and multiple discrete echogenic foci characteristic for thymic tissue are demonstrated.

Recognition of aberrant or ectopic thymus is important because these entities usually do not require therapy or surgery, except in circumstances where there is evidence of airway obstruction.

*Important: **Intrathyroid thymic tissue (Fig.3-7)**

The advances in US technology and its increasing use in the imaging evaluation of the pediatric neck and/or thyroid gland have resulted in incidental thyroid "abnormalities" commonly nowadays encountered.

Solid lesion with unusual or irregular shape, with less or more well-defined margins, with a usually parallel to the long axis of the thyroid lobe orientation.

Echogenicity heterogeneous due to internal echoes, or relatively homogeneous. But: the echogenicity of the lesion similar to that of the normal thymus.

Sometimes, the intrathyroid lesion appears to extend to the periphery or even beyond the limits of the thyroid into the adjacent soft tissues.

*DD:* The presence of hyperechoic punctate foci mimics calcification foci and may be misinterpreted as a sign of malignancy.

**Accessory thymic lobe/lobes (Fig. 5-8)**
The thymus gland is in normal position. An accessory thymic lobe extends upward from the right/left upper thymic pole to the ipsilateral lower thyroid pole, sometimes embedded in it.

**Arrest in migration (Fig. 9-12)**

The thymus gland is not depicted in anterior mediastinum. A solid mass, with echotexture similar to that of thymic tissue is found in the lateral neck, anywhere from the mandible to the base of the neck.

Another uncommon clinical condition and sonographic appearance: **Superior cervical extension of the thymus** (Fig. 13-16)

Cervical extension of the thymus is a normal finding, appears as a soft tissue mass, isoechoic to the thymus, extending above the manubrium, anterior to the trachea and great vessels, in direct continuity to the mediastinal thymus gland. Sometimes, it is clinically intermittently apparent during crying or coughing, due to the increase of the intrathoracic pressure, causing parents' anxiety.

**US important morphological features of cervical thymic tissue (independently of its location):**

- Angulated, and/or elongated configuration, geographic distribution of thymic tissue
- It molds over adjacent anatomic structures rather than displacing or invading them.

**II) Thymopharyngeal duct remnants** (Thymic cyst, thymopharyngeal duct cyst) (Fig. 17, Fig.18, Fig. 19-22)

**Differential diagnosis of cervical cystic masses in children**

The differential diagnosis when presented with a fluid-filled cyst in the cervical region includes thymic cysts, brachial cleft cysts, thyroglossal duct cysts, dermoid cysts, thymic tumors, cystic hygromas, teratomas and abscesses.

The criteria for distinguishing a cervical thymic cyst from other cystic cervical masses such as brachial cleft cyst or lymphangioma (cystic hygroma) are: age of the patient, gender, location and features of its manifestation.

Brachial cleft cysts usually appear after adolescence,

lymphangiomas appear before the age of 2 years, whereas
the majority of the thymic cysts occur between the age of 2 and 13 years. Most authors report a male predominance of thymic cysts.

A **cervical thymic cyst** is usually a multilocular mass, occurring most frequently in the anterior cervical triangle, usually on the left side and close to the anterior border of the sternocleidomastoid muscle.

Lymphangiomas are cystic masses with linear septae that occur usually in the posterior cervical triangle, often violating normal fascial planes. They may extend into the anterior triangle of the neck and have a mediastinal extension in only 3-10% of patients while mediastinal extension occurs in 50% of cervical thymic cysts.

Brachial cleft cysts tend to occur more commonly in the upper half of the neck and develop from tissue of the tonsilar fossa descending between the internal and external carotid arteries,

whereas thymic cysts tend to occur more frequently in the lower half of the neck and are located anteriorly to sternocleidomastoid muscle and into the carotid sheath, between the common carotid artery and the vagus nerve.

Thymic cysts, frequently, adhere to other surrounding structures such as the jugular vein, the phrenic nerve, the recurrent laryngeal nerve and the thyroid gland.

Brachial cleft cysts do not enlarge with Valsalva maneuver, rarely extend to the clavicle and more often occur along with signs and symptoms of recurrent infection.

On the contrary, clinically, thymic cysts usually transilluminate and expand with a Valsalva maneuver.

There may be intralesional echoes on US with or without superimposed hemorrhage or infection.
Fig. 1

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Fig. 2: Bilateral aberrant thymic tissue

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Fig. 3: Ectopic intrathyroid thymic tissue, with punctate echogenic foci

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**Fig. 4:** Ectopic intrathyroid thymic tissue, more hypoechoic, compared to Fig.3

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**Fig. 5:** Ectopic intrathyroid thymic tissue (Fig 6, 7 same case)

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**Fig. 6:** Ectopic intrathyroid thymic tissue and accessory thymic lobe (same case Fig. 5, Fig. 7)

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**Fig. 7:** Magnified view, Fig. 6

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Fig. 8: Accessory thymic lobe, between left thyroid lobe and normotopic left thymic lobe

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**Fig. 9:** Fig.9-12: Arrest of migration of left thymic lobe, lying along the left carotid artery. Right thymic lobe in its normal position.

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**Fig. 10:** Fig.9-12: Arrest of migration of left thymic lobe, lying along the left carotid artery. Right thymic lobe in its normal position.

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**Fig. 11:** Fig.9-12: Arrest of migration of left thymic lobe, lying along the left carotid artery. Right thymic lobe in its normal position

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Fig. 12: Fig.9-12: Arrest of migration of left thymic lobe, lying along the left carotid artery. Right thymic lobe in its normal position

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**Fig. 13:** Fig.13-16. Superior herniation of thymus gland during coughing. Fig. 13. At rest.

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Fig. 14: Fig.13-16. Superior herniation of thymus gland during coughing. Fig. 14. Coughing

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Fig. 15: Fig. 13-16. Superior herniation of thymus gland during coughing. Fig. 15. At rest.

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Fig. 16: Fig. 13-16. Superior herniation of thymus gland during coughing. Fig. 16. Herniation of the left thymic lobe during coughing.

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Fig. 17: Thymic cyst with internal echoes and peripheral thymic tissue with characteristic tiny punctate foci.

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Fig. 18: Thymic cyst in continuity with the normotopic thymic gland.

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Fig. 19: Fig. 19-22. Persistent left lower thymopharyngeal duct (cystic dilatation) Fig.19. Longitudinal section of left thyroid lobe.

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**Fig. 20:** Fig. 19-22. Persistent left lower thymopharyngeal duct (cystic dilatation) Fig.20. Longitudinal section of persistent left thymopharyngeal duct with echogenic fluid, adjacent to the left thymic lobe.

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**Fig. 21:** Fig. 19-22. Persistent left lower thymopharyngeal duct (cystic dilatation) Fig. 21. Transverse view. Cystic dilatation of left thymopharyngeal duct. Note its close relationship with the left common carotid artery and internal jugular vein.

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**Fig. 22:** Fig. 19-22. Persistent left lower thymopharyngeal duct (cystic dilatation)

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Conclusion

The sonographic examination is ideal for the recognition of cervical thymic tissue and its pathology.

A thorough knowledge of:

- the embryology and anatomy of the thymus,
- normal variations and ectopic locations of the thymus and its congenital abnormalities,

is essential to avoid diagnostic errors and clinical dilemmas, or to determine proper management.

According to our experience and recent literature data, it is possible to diagnose cervical thymic tissue by sonography in the majority of cases, and there is no necessity for biopsy or surgical excision when the imaging diagnosis is made.
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Personal Information

Vakaki Marina
Pediatric Radiologist
Department of Radiology
Children's Hospital "P & A.Kyriakou", Athens, Greece
e-mail: geopmar@gmail.com