

Chapter 74 – Thyroglossal Duct Cyst

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A thyroglossal duct cyst (TGDC) is the most common mass encountered in the midline of the upper part of the neck. Although a TGDC may develop at any location along the thyroglossal duct tract, it is most commonly manifested as an upper midline cervical mass, usually adjacent to the hyoid bone and thyrohyoid membrane. TGDC may also be found in the submental space or as a low cervical mass.^[1] Because thyroid development occurs along the midline of the neck, the cyst rarely occurs in a lateral position. Recently, however, six cases of intrathyroid TGDC have been reported in the literature (two children^[2] and four adults^[3,4]).

Successful management of thyroglossal duct remnants must be based on an understanding of the embryology and developmental anatomy of the thyroid gland.^[5] The thyroid anlage appears in the 2.0- to 2.5-mm embryo as a midline structure and projects downward from the floor of the pharynx at a point that corresponds to the foramen cecum in adults.^[6] During the course of development, the thyroid descends along the midline to its position in the anterior aspect of the neck, where it forms a tract known as the thyroglossal duct and maintains an attachment to the base of the tongue. The hyoid bone divides this tract into upper and lower segments during the second embryologic month, when the thyroid gland assumes its final position anterior to the trachea in the root of the neck, at which time the tract is resorbed. The hyoid bone arises from the second branchial arch and grows forward after descent of the thyroid. Rotation of the hyoid during its development results in the thyroglossal duct being drawn posteriorly and cranially at the inferior margin of the bone. Fusion of the second branchial arch anlage in the midline may result in entrapment of the thyroglossal duct by the hyoid bone. Dr. Sistrunk, who understood the developmental anatomy, included removal of the body of the hyoid as a fundamental step in the prevention of recurrence. Failure of the resorption process, or the presence of retained epithelial rests along the tract, forms the basis for the development of a TGDC. No communication occurs between the thyroglossal duct remnant and the skin because of the absence of embryologic communication in some branchial cleft abnormalities.

The differential diagnosis of a midline cervical mass includes

- Thyroglossal duct cyst
- Dermoid cyst
- Lipoma
- Hemangioma
- Fibroma
- Cervical lymphadenopathy

In a series of 1316 cases reported in the literature, 31% of the patients were younger than 10 years, 20.4% were in their second decade of life, 13.5% were in their third decade, and 34.6% were older than 30 years.^[7] We reviewed the results of the University of Pittsburgh TGDCs in adults and identified 45 patients older than 18 years in a group of 104 patients.^[8] TGDCs occur equally in both genders as demonstrated by Allard in a series of 1747 patients with TGDCs collected from the literature,^[7] in which 870 were male and 877 were female. Although most develop in childhood, a TGDC may not become clinically apparent until well into adulthood. Approximately 0.6% of such cysts are found in patients older than 60 years,^[9] and only 8 cases of TGDC have been reported in patients 70 years or older.^[10–12] Though unusual, cancer involving TGDCs has been reported.^[13] These authors recommended removal of all suspected TGDCs in the elderly because of the notable rate of malignant transformation in their series. Fine-needle aspiration biopsy (FNAB) may be helpful in identifying cancer preoperatively in this population.

PATIENT SELECTION

Surgical excision of a mass in the midline of the neck is performed for diagnostic and therapeutic purposes. The following are indications for excision of what appears to be a TGDC:

- The presence of an undiagnosed mass
- Cosmetic appearance
- A history of infection in the mass
- The possibility of cancer originating in a TGDC

On initial evaluation, TGDCs may be relatively small or very large (Fig. 74-1). Some patients have a history of current or recurrent infection. In some cases, incision and drainage, packing, and antibiotics have been used to treat an abscess in this area without the treating physician recognizing that it is an infection in a TGDC. In these circumstances, recurrent infection is to be expected. Patients may also have a history of chronic drainage from the mass if previous incision plus drainage has been performed or an attempt that is less extensive than a Sistrunk procedure has been made to remove a TGDC.

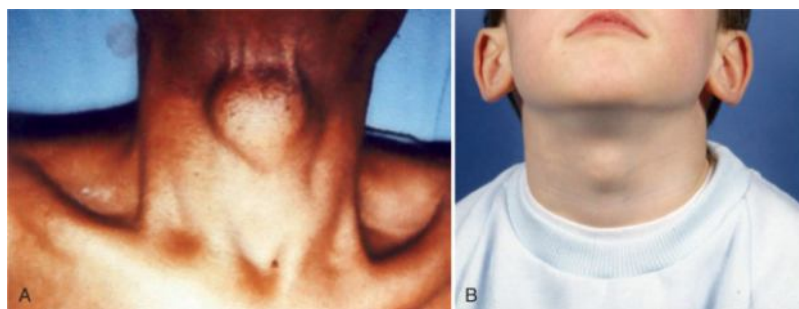


Figure 74-1 Thyroglossal duct cysts may be large (A) or small (B) on initial evaluation.

Carcinoma arising in a thyroglossal duct remnant was first reported in 1925 by Ashurst and White.^[14] Since that time more than 100 such cases have been reported.^[7] Rampaul and coauthors found 160 cases published in the literature in the past 85 years.^[15] Patients in these series range in age from 6 to 91 years, with an average age of 39.2 years. Most of the patients were in the third through the sixth decades of life. Cancer occurs more frequently in females than in males. The diagnosis of carcinoma is seldom made definitively preoperatively; however, the presence of calcification in a TGDC on imaging studies suggests papillary carcinoma (Fig. 74-2) because these areas of calcification are psammoma bodies, a histologic hallmark of papillary carcinoma. The first case of a benign TGDC with psammoma bodies was reported in a 3-year-old patient.^[16] It has been suggested that the presence of calcification in association with a TGDC may be a specific radiologic marker for carcinoma.^[17] The diagnosis of carcinoma arising in a TGDC is usually made histologically after the mass has been excised. Pathologic diagnoses in these cases include papillary adenocarcinoma, follicular adenocarcinoma, mixed papillary and follicular adenocarcinoma, adenocarcinoma, and squamous cell carcinoma.^[7]

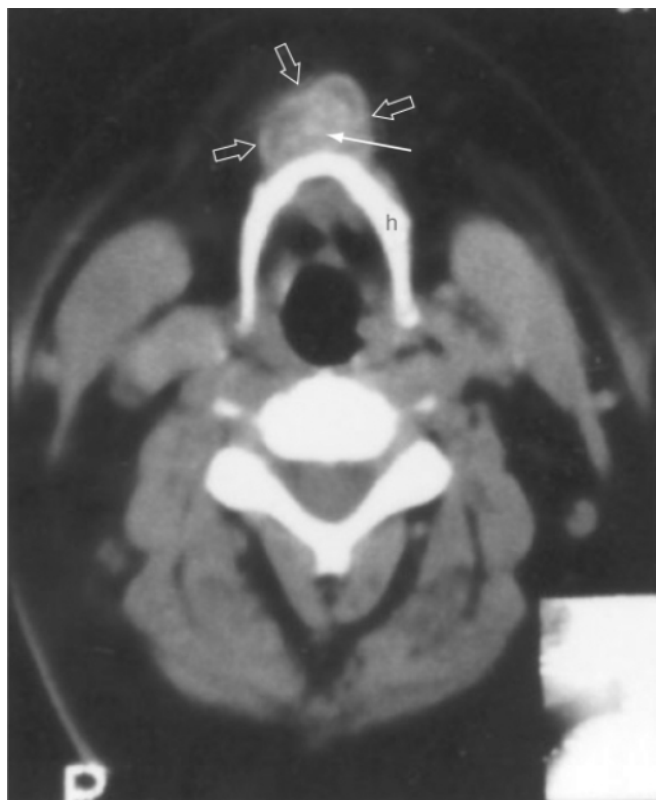


Figure 74-2 Calcification in an imaging study of a thyroglossal duct cyst raises suspicion of papillary carcinoma arising in a thyroglossal duct cyst. h, hyoid bone; long arrow, calcification; open arrows, thyroglossal duct cyst.

Management of carcinoma arising in a TGDC remains somewhat controversial because this problem is encountered relatively infrequently (<1%). When the diagnosis of carcinoma is made, a radionuclide scan with radioactive iodine (^{131}I) is performed. If the thyroid gland appears normal, thyroidectomy is not necessary. Suppression of thyroid-stimulating hormone with thyroid hormone is initiated, along with systematic follow-up and serial determination of thyroglobulin levels. Resection of a TGDC by the Sistrunk procedure has been demonstrated to be oncologically adequate in the management of differentiated thyroid carcinoma in the absence of extracapsular extension, lymph node metastasis, or thyroid abnormality. If a cold nodule is found in the thyroid gland, thyroidectomy should be performed with central compartment nodal dissection. Neck dissection is indicated if cervical metastasis is identified. Ultrasound-guided fine-needle aspiration may also be useful in the setting of possible cancer in a TGDC.

TGDCs can produce a variety of symptoms ranging from the mere presence of the mass to dysphagia, coughing, a choking sensation, or upper respiratory obstruction. Airway obstruction is a serious, though rare complication that may occur as a result of involvement of the preepiglottic space. Lübben and colleagues reported a 62-year-old man with progressive respiratory obstruction from a $5 \times 3 \times 2$ -cm bilobed mass that extended through the preepiglottic space into the larynx.^[18] The authors also noted that three cases of invasion of the larynx with a TGDC had been reported by Slotnik and associates.^[19] Brown and coauthors reported a 37-year-old patient with complaints of a sore throat, a choking sensation, and difficulty breathing while recumbent.^[20] Invasion of the anterior larynx by a TGDC filling the preepiglottic space was found on computed tomography (CT). No patient in our series experienced respiratory obstruction.^[8]

TGDCs are, on average, 1 to 2 cm in size but may range up to 10 cm.^[7] The mass is nontender unless it is infected. The skin will be freely movable over the cyst unless the cyst has been or is currently infected. Characteristically, the mass moves with swallowing or protrusion of the tongue, which reflects attachment to the hyoid bone. If the patient has a midline mass in the neck that does not move with swallowing, a pathologic entity other than a TGDC should be considered. If there is no history of infection in the mass, it is often possible for the examiner to confirm the diagnosis via palpation of the thyroglossal duct by grasping the mass between the thumb and forefinger and reaching toward the hyoid bone (Fig. 74-3).



Figure 74-3 The thyroglossal duct may be palpated deep to the cyst.

PREOPERATIVE PLANNING

The history and physical examination have remained the mainstay for diagnosis and therapeutic decision making relative to TGDCs. However, a number of studies have been proposed to aid in the diagnosis of a midline neck mass:

- Ultrasonography
- High-resolution CT
- Magnetic resonance imaging (MRI)
- Radionuclide scanning
- FNAB

Radionuclide studies are helpful in identifying functional thyroid tissue and distinguishing it from other nonthyroid abnormalities. Radioisotopic scanning identifies functioning thyroid tissue in a TGDC in approximately a third of patients. Thyroid scanning is reliable in confirming the presence of functioning thyroid tissue outside the TGDC itself. The scan, therefore, is not done specifically to diagnose a TGDC but is used to eliminate the possibility of mistaking an ectopic thyroid gland for a TGDC. Advocates of routine preoperative radioactive scanning of the thyroid gland in children with suspected TGDCs note the substantial frequency of ectopic thyroid tissue manifested as a mass in the midline of the neck in the absence of a functioning thyroid gland lower in the neck.^[21] Excision of a misdiagnosed ectopic thyroid gland will result in postoperative hypothyroidism. The literature contains reports of myxedema after inadvertent excision of a mass in the neck that proved to be the only functioning thyroid tissue. Radkowski and coworkers^[22] reported five of eight patients with an ectopic thyroid or thyroid nodule in their review who had a preoperative history suggestive of hypothyroidism. They suggest that a careful history and physical examination to establish whether there are symptoms suggestive of hypothyroidism, including fatigue, chronic constipation, developmental and growth delay, and excessive somnolence on waking, will aid in identifying patients with ectopic thyroid tissue who are at risk for the development of postoperative myxedema. If there is suspicion of hypothyroidism, they suggest that thyroid function tests be performed. Any abnormality on these tests should prompt a preoperative nuclear scan to rule out ectopic thyroid tissue. The authors present an algorithm for the management of anterior neck swelling (Fig. 74-4).

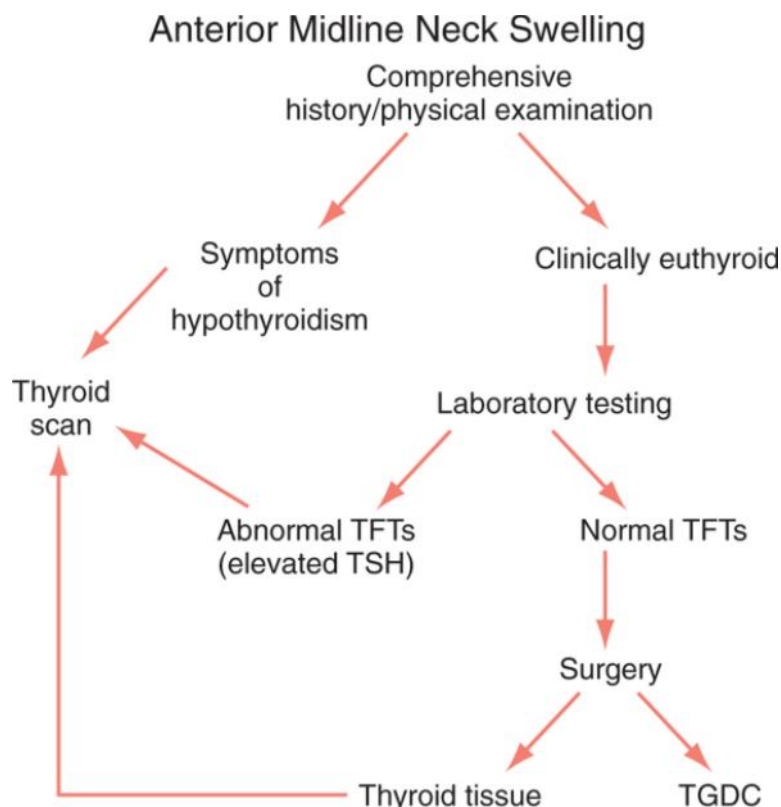


Figure 74-4 Algorithm for evaluation of a midline neck mass. TFTs, thyroid function tests; TGDC, thyroglossal duct cyst; TSH, thyroid-stimulating hormone.

Ahuja and associates^[23] recommend high-resolution ultrasound for ideal initial investigation, especially in children, because it does not require ionizing radiation or sedation, is readily available and inexpensive, and provides the surgeon with the necessary preoperative information. The authors studied 23 children with TGDCs via ultrasound and concluded that TGDCs in children are not simple cysts but have a complex pattern, including anechoic (13%), pseudosolid (56.5%), and heterogeneous (30.5%) areas.^[23]

The success of CT and MRI in detecting TGDCs is also well documented.^[24,25] King and coworkers^[26] investigated the utility of MRI in 16 patients with TGDC via T1, T2, and T2 fat saturation images. Contrast-enhanced images were obtained in five patients. They concluded that TGDCs most commonly have high T1 signal intensity because of the high protein content in the fluid. The tract leading to the base of the tongue is infrequently seen, the diagnosis being determined by the intimate relationship to the hyoid bone and strap muscles. Intralaryngeal extension of TGDCs was noted more frequently in adult patients than had been reported previously.

FNAB has gained popularity in recent years because it is a simple, rapid, and inexpensive technique with low complication rates.^[27] FNAB is not used routinely in the diagnosis of TGDC. Although the diagnostic criteria for TGDC and papillary carcinoma have been well defined, FNAB fails to achieve an accurate preoperative diagnosis of papillary carcinoma in TGDCs.

Astl and coauthors^[28] described their experience with TGDCs and cancer of the thyroid in three patients and reviewed the literature. They used clinical and ultrasound examination and FNAB for preoperative assessment. Diagnostic and therapeutic procedures included both thyroglossal duct remnant excision (Sistrunk or Schlange procedure) and total thyroidectomy. Postoperative radiation therapy or radioiodine ablation was used in cases of carcinoma arising in a thyroglossal duct remnant or carcinoma of the thyroid associated with a thyroglossal duct remnant. Although the therapeutic approach could be a matter of discussion, most patients agreed with their suggestion of relatively radical but nonmutating treatment.

On the other hand, Patel and colleagues^[29] reviewed 57 evaluable patients with well-differentiated thyroid carcinoma arising in a TGDC who were reported in the recent literature, as well as 5 cases of their own. A Sistrunk procedure was performed for the TGDC in the majority (90%) of patients. Papillary carcinoma was the most frequent (92%) histologic type. Total thyroidectomy was performed after the diagnosis of TGDC carcinoma in approximately half of the 62 patients. A malignant tumor was found in 27% of the thyroidectomy specimens. Postoperative ¹³¹I was administered to 16 (26%) of the patients. There were no disease-related deaths reported in any of these patients. Univariate analysis revealed that the only predictor of overall disease-free survival was the extent of primary surgery for the TGDC. The addition of total thyroidectomy to the Sistrunk procedure did not have a significant impact on outcome. The Sistrunk procedure is adequate for most patients with incidentally diagnosed TGDC carcinoma in the presence of a clinically and radiologically normal thyroid gland. The results of the Sistrunk operation with adequate excision are excellent, and the concept of risk groups should be used to identify patients who would benefit from more aggressive treatment.

DESCRIPTION OF SURGERY

In 1893, Schlange described a procedure that included removal of the midportion of the hyoid bone in continuity with the cyst.^[30] The Sistrunk procedure extended the Schlange procedure by also including resection of a portion of the base of the tongue and has been the standard surgical approach for TGDCs since it was first described in 1920.^[31] The Sistrunk procedure has achieved excellent results, with recurrence rates between 1.5% and 4%.^[32] The operation is based on precise knowledge of the embryology and developmental anatomy of the thyroglossal duct. The operation includes excision of the cyst in continuity with the midportion of the body of the hyoid bone and a block of muscle that theoretically includes the thyroglossal duct remnant dissected up to its origin in the area of the foramen cecum.

Brereton and Symonds^[32] reported a 38% recurrence rate in 50 patients in whom the hyoid bone was not resected or the duct above the hyoid bone was dissected with an inadequate margin of surrounding muscle. Although excellent results with the Sistrunk procedure have been obtained, it is clear that troublesome recurrences are still encountered. Marianowski and coauthors^[6] reported a series of 74 patients who underwent a Sistrunk procedure for the preoperative diagnosis of TGDC. Fifty-seven were confirmed histologically as TGDCs. Recurrence was noted in 15% of confirmed TGDCs. Four individual risk factors have been identified: number of infections before surgery, previous surgical procedure, age (<2 years), and multicystic lesion on histopathology.

The operation is carried out under general anesthesia with the patient in the supine position. A folded blanket, or a towel in younger children, is then placed beneath the shoulders to extend the head. The transverse incision is placed in the midline in the major skin crease in the upper part of the neck to achieve a fine scar (Fig. 74-5A).

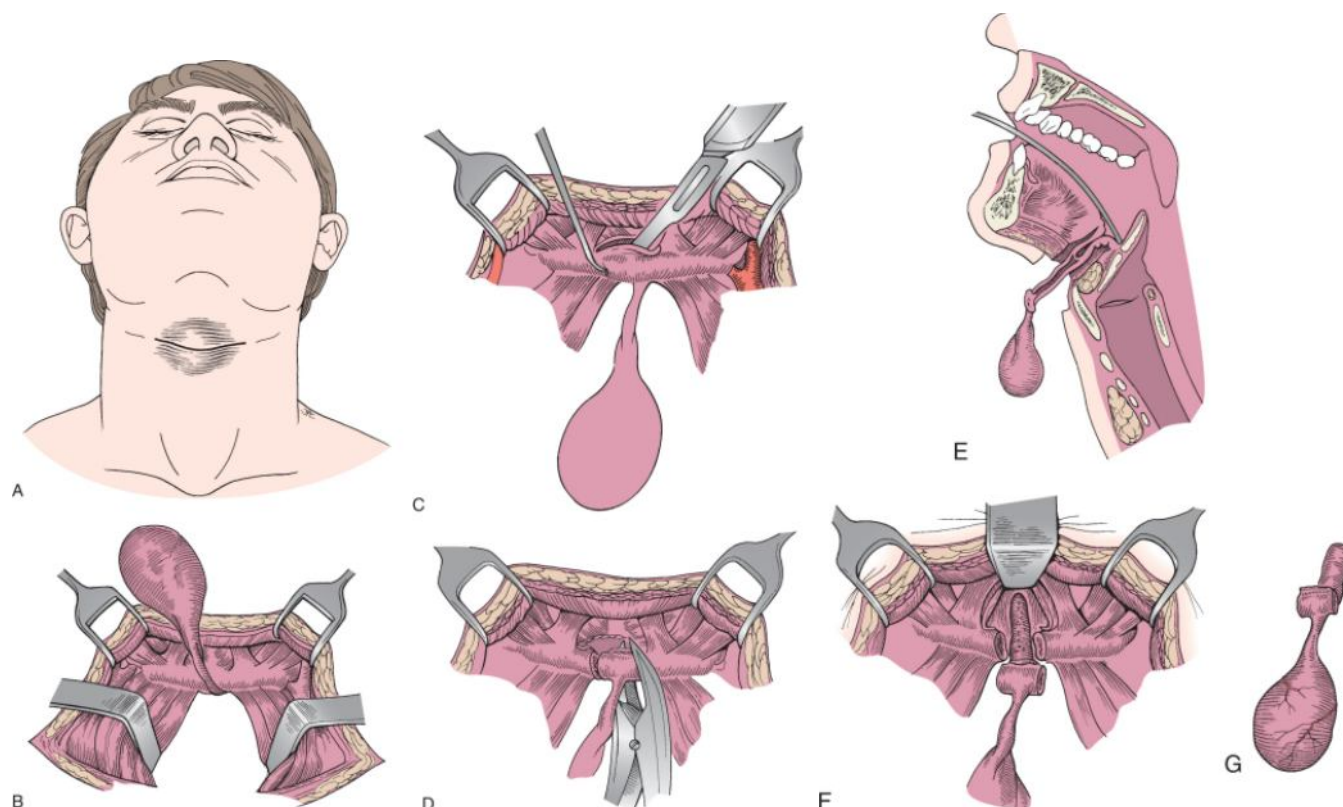


Figure 74-5 A, The incision is placed in the major skin crease in the midline of the neck. B, The cyst is identified, the strap muscles are retracted, and the cyst is pedicled on the hyoid bone. C, The hyoid bone is grasped with a tenaculum and skeletonized. D, The hyoid bone is transected with a bone cutter. E, A curved retractor placed in the oral cavity brings the tongue base into the wound, thereby facilitating dissection of the thyroglossal duct. F, The retrohyoid portion of the duct is excised together with a block of muscle (duct) tissue, up to and including the tongue base as far superior as the foramen cecum. G, The resected specimen must include the cyst, the midportion of the body of the hyoid bone, and a core of the muscle of the tongue base.

If a draining sinus is present or if the cyst has previously been infected and the skin is adherent to the cyst, an elliptical incision is made such that the sinus tract or involved skin is removed with the specimen. The incision is carried down through fat and subcutaneous tissue. The inferior flap does not usually require much elevation, whereas the flap superior to the hyoid bone must be elevated to allow adequate resection. The strap muscles are identified, an incision is made to facilitate separation of the strap muscles, which may already be separated by the cyst, and blunt retractors are used to retract the strap muscles. Via sharp and blunt dissection, the cyst is then dissected free of the thyroid cartilage and thyrohyoid membrane and pedicled on the hyoid bone superiorly (see Fig. 74-5B). In patients who have not had infection, the planes are easily identified, thus making dissection straightforward. However, in patients with previous infection, tissue planes are obliterated and bleeding may be more pronounced. Keeping dissection close to the cyst in the midline will avoid injury to vital structures and ensure that the entire cyst is removed.

The hyoid bone is then grasped with a tenaculum. Using sharp dissection with a scalpel or a sharp elevator, the muscle and soft tissues are dissected from the body of the hyoid bone. Sharp dissection should not be carried out superior or inferior to the hyoid bone to avoid transecting the thyroglossal duct (see Fig. 74-5C). After skeletonizing the body of the hyoid bone, a small sharp bone cutter is used to transect the hyoid bone (see Fig. 74-5D). Forceps are then placed on the hyoid bone and the bone elevated anteriorly. A curved retractor (baby Deaver) is next placed transorally such that the end of the retractor is in the vallecula. Pressure applied to the retractor displaces the tongue base into the wound, thus enabling accurate dissection of the suprahyoid musculature (see Fig. 74-5E). The retrohyoid portion of the duct is excised together with an ellipse of tissue up to the base of the tongue, including the foramen cecum (see Fig. 74-5F). If the vallecula is inadvertently entered during the dissection, the defect should be closed with interrupted nonabsorbable suture.

Copious irrigation is applied, particularly if the oropharynx has been entered. A Hemovac drain is inserted 1 cm lateral to one edge of the incision, and the wound is closed in layers by approximating the strap muscles in the midline and then the subcutaneous tissue. The skin incision is closed with 6-0 fast-absorbing suture. An adhesive is then applied, and the incision is covered with Steri-Strips, followed by a sterile pressure dressing.

The resected specimen should consist of the cyst, the midportion of the body of the hyoid bone, and a generous core of muscle from the base of the tongue removed en bloc with or without the mucosa of the area of the foramen cecum (see Fig. 74-5G). Pathologic evaluation may reveal simple or respiratory columnar epithelium as the majority of the specimen; however, squamous epithelium is another predominant pattern, and 20% of specimens contain thyroid tissue. Waddell and colleagues^[33] studied the pathology of 61 patients who had a neck mass clinically diagnosed as a TGDC. Their study revealed that although the majority of the specimens contained thyroglossal duct cyst remnants, only approximately half could be described as a true cyst. Normal thyroid tissue is present in approximately 25% of duct remnants. The authors suggest that it might be more accurate if the term "thyroglossal tract remnants" were used to cover this whole group of conditions (Table 74-1).

Table 74-1 -- NUMBER OF SPECIMENS IN EACH HISTOLOGIC GROUP

	n	%
Cyst	28	46
Duct (without evidence of a cyst)	24	39
Fibrous tract	5	8
Abscess cavity	2	3
Lymph node	1	2
Epidermoid cyst	1	2

Reprinted with permission from Waddell A, Saleh H, Robertson N, et al: Thyroglossal duct remnants. *J Laryngol Otol* 114:128-129, 2000.

POSTOPERATIVE MANAGEMENT

The Hemovac drain is removed when drainage amounts to less than 10 mL of serum, which ordinarily occurs within 24 to 48 hours. The outer dressing is removed, and the patient is discharged from the hospital. Antibiotics are not generally used unless the oral cavity has been entered or there has been a history of infection or abscess formation preoperatively. Contamination of the surgical field by the cyst contents is an indication for the administration of antibiotics.^[34] Perioperative antibiotics are administered for only 24 hours. The patients in our series and others^[34] were generally discharged on the first postoperative day.

COMPLICATIONS

Complications are unusual and may consist of postoperative hematoma, seroma, or wound infection. Hematoma is treated by returning the patient to the operating room to evacuate the hematoma and ensure hemostasis. Airway distress has not been encountered.

Hoffman and Schuster studied the results of 90 patients treated at the Children's Hospital in Boston.^[5] The authors concluded that the best opportunity for curative excision is at the initial surgery. Previous inadequate or unsuccessful excision serves as a major risk factor for recurrence. This was reflected in the 30% recurrence rate after radical re-excision in patients referred to Children's Hospital of Boston for further management of recurrence, which contrasted with the recurrence rate of 6.2% in patients undergoing primary excision by the authors. The authors' second conclusion was that there does not seem to be any factual data regarding the absolute incidence of a TGDC traversing the central portion of the bone. Although the Sistrunk procedure included the hyoid bone because of embryologic evidence that the duct traversed the hyoid bone, this may not necessarily be so. Hoffman and Schuster reported that dissection of soft tissue above the hyoid bone (Fig. 74-6) is important because of the presence of accessory ducts and diverticula from the main duct and pharyngeal mucosa. They suggest wide dissection of a generous core of muscle between the superior aspect of the hyoid bone and the base of the tongue. Variability in the anatomy of the duct was noted in 7.8% of their 90 patients. They also pointed out that the best management for recurrent disease is wide secondary excision with removal of an additional portion of the hyoid bone or suprahyoid soft tissue as appropriate. Ducic and colleagues^[35] indicated that when a branched TGDC occurs, residual tracts can be left behind while removing the main tract. Multicystic tracts on histology were more commonly noted in patients with recurrent TGDCs. All recurrences were managed successfully by wider excision. They also found a correlation between recurrence after the Sistrunk procedure and a history of recent (6 month) infection of the cyst. Arborization of the TGDC at the level of the hyoid bone occurred at a much higher rate in patients who failed a Sistrunk procedure than in the group that was cured by a Sistrunk procedure.



Figure 74-6 Longitudinal section through the thyroglossal duct demonstrating the tract (A), as well as multiple diverticula (B). Also seen are serous mucous glands (C) well away from main duct and adjacent to striated musculature (D) that was taken along with duct. (Reprinted with permission from Hoffman MA, Schuster SR: *Thyroglossal duct remnants in infants and children: Reevaluation of histopathology and methods for resection.* *Ann Otol Rhinol Laryngol* 97:484, 1988.)

Patients with recurring TGDCs have recurrent swelling and tenderness and discoloration of the skin in the area of the previously operated TGDC (Fig. 74-7). Alternatively, these patients may have a cutaneous tract with drainage. It may not necessarily be in the midline but may be in another area such as the submental or submandibular space. An elliptical excision of the skin surrounding the fistula tract is then required, followed by dissection carried from the fistula to the area of the hyoid bone. Our experience is that patients with a recurrent TGDC have not usually had the body of the hyoid bone removed. In cases in which the cyst recurs after the hyoid bone has been already excised, a core of tissue, as previously described, is removed up to and including the base of the tongue to incorporate multiple tracts.



Figure 74-7 A and B, Recurrent thyroglossal duct cysts.

PEARLS

- Preoperative ultrasonography provides valuable information regarding whether the thyroid gland is present when a TGDC is found. This procedure is particularly recommended in children because it spares them exposure to radiation.
- Removal of the TGDC, the midportion of hyoid bone, and a portion of the base of the tongue (Sistrunk procedure) remains the procedure of choice because of the low rate of complications or recurrence.
- Cancer arising in a TGDC is often an unexpected finding when a TGDC is removed. The presence of calcifications on preoperative scanning is suggestive but not diagnostic of cancer.
- A ^{131}I study should be carried out when the diagnosis of cancer in the TGDC is made pathologically. If no abnormalities are noted in the thyroid gland, no further surgery is indicated.

PITFALLS

- Failure to conduct a preoperative evaluation may result in inadvertent removal of the only functioning thyroid gland.
- Failure to remove the midportion of the body of the hyoid bone and sufficient soft tissue at the base of the tongue results in a high rate of recurrence of TGDCs.
- Entering the pharynx during the procedure may result in postoperative infection.
- Unless the skin incision is placed in the natural skin crease overlying the TGDC, an unsightly scar may result.
- Carcinoma may arise in a TGDC, even in children, so removal of TGDCs should not be delayed.