*Cite as: Delantoni A, Onder M, Orhan K: B-mode and color Doppler imaging of different types of branchial cleft cysts in children. A multicenter study and review of the literature. J Ultrason 2022; 22: e174–e178. doi: 10.15557/JoU.2022.0028.*

**Submitted:** 20.11.2021
**Accepted:** 25.02.2022
**Published:** 11.07.2022

# B-mode and color Doppler imaging of different types of branchial cleft cysts in children. A multicenter study and review of the literature

Antigone Delantoni[1], Merve Onder[2], Kaan Orhan[2]

[1] *Department of Oral Surgery, Implant Surgery, and Radiology, Faculty of Dentistry, Aristotle University of Thessaloniki, Greece*
[2] *Faculty of Dentistry, Department of Dentomaxillofacial Radiology, Ankara, Turkey*

*Correspondence: Dr. Kaan Orhan, Ankara University, Faculty of Dentistry, Department of Dentomaxillofacial Radiology, Besevler, Ankara Turkey 06500; tel.: 00 90 (535) 6765010, fax: 00 90 (312) 2123954, e-mail: call53@yahoo.com*

DOI: 10.15557/JoU.2022.0028

**Keywords**
branchial cleft cyst; Doppler ultrasonography; cervical lymphoepithelial cyst; branchial anomaly

**Abstract**

**Aim:** The term "branchial cleft cyst" refers to the lesions that can be considered synonymous with cervical lymphoepithelial cysts. Although relatively rare, they constitute the second major cause of head and neck pathologies in childhood. This study aimed to report the clinical presentations, diagnosis, and management of pediatric patients with the pathological diagnosis of branchial cleft cyst. **Material and methods:** This study was a retrospective analysis of the records of 33 patients with the diagnosis of branchial cyst, in two different university hospitals, in two different populations. **Results:** Thirty-three cases of branchial cleft cysts were seen in 33 patients: 17 females and 16 males. The majority (16 patients) were 2nd branchial cleft cysts. Accurate diagnosis of branchial cleft malformation was made via imaging in 20 of the 21 (95%) patients that underwent preoperative surgical ultrasonographic imaging. **Conclusion:** Branchial cleft cysts are frequently incorrectly diagnosed and ignored in the differential diagnosis. Thus, the diagnosis is often delayed, resulting in the mismanagement of affected patients. A branchial cyst should be suspected in any patient with a swelling in the lateral aspect of the neck, regardless of whether the swelling is solid or cystic, painful or painless. The use of ultrasonography can dramatically help clinicians with distinguishing branchial cleft cysts from other similar lesions of the head and neck.

## Introduction

The term "branchial cleft cyst" (BCC) refers to lesions which can be considered synonymous with cervical lymphoepithelial cysts. Although relatively rare, they are the second major cause of head and neck pathologies in childhood. They were reported in the literature as far back as 1930[(1)], though the first analytic paper was published in 1972[(2)]. Of the congenital masses related to embryonic remnants, approximately 70% are thyroglossal duct sinuses and cysts, 25% are branchial cysts and sinuses, and 5% are cystic hydromas[(3–4)]. Branchial cleft cysts are located unilaterally in the side of the neck and are considered to originate from either incomplete fusion of a branchial cleft resulting in the creation and formation of a cyst (75% of cases) or a residual cavity (the remaining 25%). Branchial cleft cysts form during the development of the embryo when tissues in the neck (branchial cleft) fail to develop normally[(5–8)]. The birth defect may appear as open spaces, called cleft sinuses, which may develop on one or both sides of the neck. A branchial cleft cyst may form from fluid drained from a sinus. The cyst or sinus can become infected, and there is also a possibility of recurrent inflammations of the lesion[(9–11)]. The principles of patient management include early diagnosis, monitoring of the infection status, and complete excision without facial nerve injury[(12–13)].

This study aimed to report the clinical presentations, diagnosis, and management of pediatric patients with the pathological diagnosis of branchial cleft cyst.

**Tab. 1.** *1st cleft cyst: in the inferior pole of the parotid gland one can observe a cystic-like lesion of multilobular borders, septa that are of low vascularity on color Doppler, and thick content, attached to the lateral borders*

| Type | Total number | Male to female ratio | Density | Doppler ultrasound (internal or peripheral vasculature increase/no flow) |
|---|---|---|---|---|
| 1st cleft | 9 | 5/4 | All hypoechoic | 2/7 |
| 2nd cleft | 16 | 7/9 | Hypoechoic/hyperechoic (1 case)/mixed (1 case) | 1 (1 inconclusive)/14 |
| 3rd cleft | 3 | 2/1 | All hypoechoic | 0/3 |
| 4th cleft | 2 | 1/1 | All hypoechoic | 0/2 |
| Inconclusive | 3 | 1/2 | All hypoechoic | 0/3 |

## Materials and methods

A total of 33 children (16 male, 17 female) aged between 6 and 14 years, who underwent surgery for 33 branchial anomalies during the review period from 2017 to 2021, were studied retrospectively. The images were obtained from the Aristotle University of Thessaloniki School of Dentistry and the AHEPA University Hospital and Ankara University Dental Hospital.

The patients' records were extracted from retrospective digital patients' clinical records. The patients' presenting history and physical findings, as well as their preoperative imaging data, were examined. Histology results were also reviewed to ensure that the diagnosis was correct and corresponded to the medical findings in each case.

The images were selected based on the final diagnosis and the imaging features of all the lesions were described. The images were studied only based on the features that were present upon ultrasonographic examination and not the findings obtained by other imaging techniques.

The age and sex of the patients were recorded, and two observers marked down the ultrasonographic features. The level of branchial cleft anomaly was identified in all cases. The first cleft anomalies were easily differentiated and distinguished from other anomalies, since they typically occur around the ear area. In contrast to the first arch clefts, second, third, and fourth branchial anomalies tend to occur lower in the neck. While these branchial anomalies are typically distinguished by their course as well as their openings (the internal opening in the pharynx and the external opening in the neck), these features may not always be present. In cases where the collated data was inconclusive, they were reflected as such.

## Results

Table 1 shows the type, female/male ratio, and Doppler ultrasound features. The B-mode ultrasound showed all cases as hypoechoic, however, Doppler scans showed "increased internal or peripheral vasculature related to lesion" in a limited number cases (three cases and one inconclusive). Regarding their distribution by sex, 48% of the cases were male whereas 52% of cases were female. The female to male ratio was 1:0.92.

Of the total of 33 branchial anomalies, nine (27%) were first branchial anomalies, 16 (48%) were second branchial anomalies, three (9%) were third branchial anomalies, and two (6%) were fourth branchial anomalies. In the cases of the third and fourth branchial arch cysts, three cases were inconclusive as to their origin (3rd or 4th arch). All the patients whose origin was undetermined presented with infections of the branchial anomalies (cystic mass or abscess) (Fig. 1, Fig. 2, Fig. 3).

All the patients with first branchial cleft anomalies presented with swelling in the postauricular region. None of them had an external sinus opening, while half of them presented with either persistent or recurrent infections of the postauricular area and even abscesses, which prompted the consideration of first branchial cleft cysts.

Ultrasonographic imaging assessment was obtained for 21 of the 33 branchial cleft anomalies that were surgically treated, while the rest of the patients underwent surgical treatment based on their clinical presentations or other imaging modalities such as CT or MRI.

Accurate diagnosis of branchial cleft malformations was made via imaging in 20 of the 21 (95%) cases that underwent preoperative ultrasonographic imaging. In the one case that ultrasonography could not provide a definite

**Tab. 2.** *Table showing the differential diagnosis according to BCC types*

| Cleft cyst | Differential diagnosis |
|---|---|
| 1st | Abscesses<br>Lymphatic malformations e.g. necrotic lymph nodes<br>Parotid mucocele<br>Benign lymphoepithelial cyst<br>Warthin tumor |
| 2nd | Lymphatic malformations<br>Nodal metastasis<br>Lymphadenopathy<br>Abscess<br>Schwannoma |
| 3rd | Lymphatic malformations<br>Abscess<br>Infrahyoid thyroglossal duct cyst<br>Nodal metastasis<br>Laryngocele<br>Thyroid ectopic nodules |
| 4th | Necrotic lymph nodes<br>Abscess<br>Cystic schwannoma<br>Thyroid colloid cyst<br>Thymic cyst<br>Lymphatic malformations |

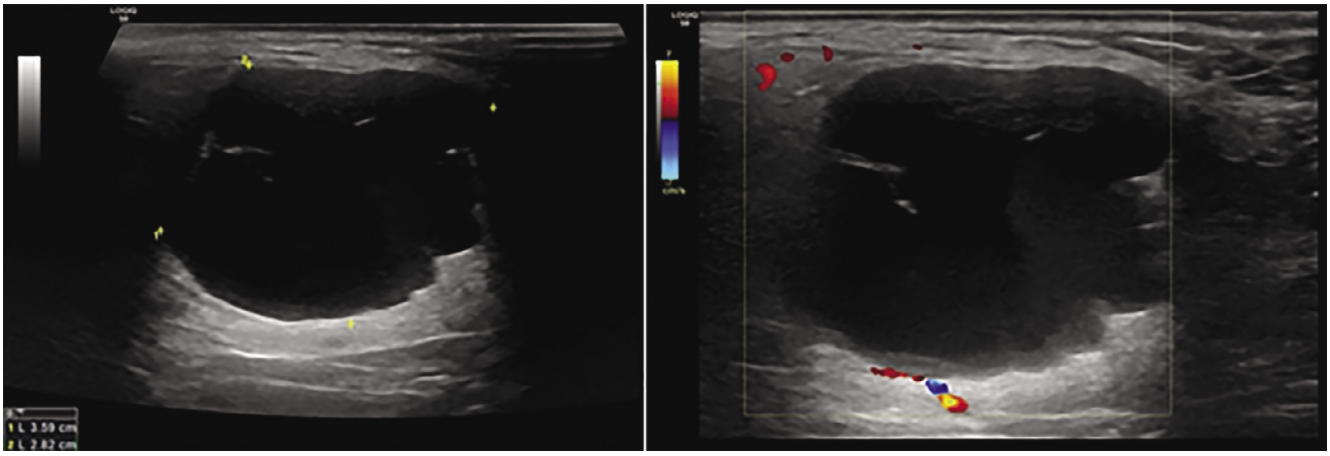

**Fig. 1.** *1st cleft cyst: in the inferior pole of the parotid gland one can observe a cystic-like lesion of multilobular borders, septa that are of low vascularity on color Doppler, and thick content, attached to the lateral borders*

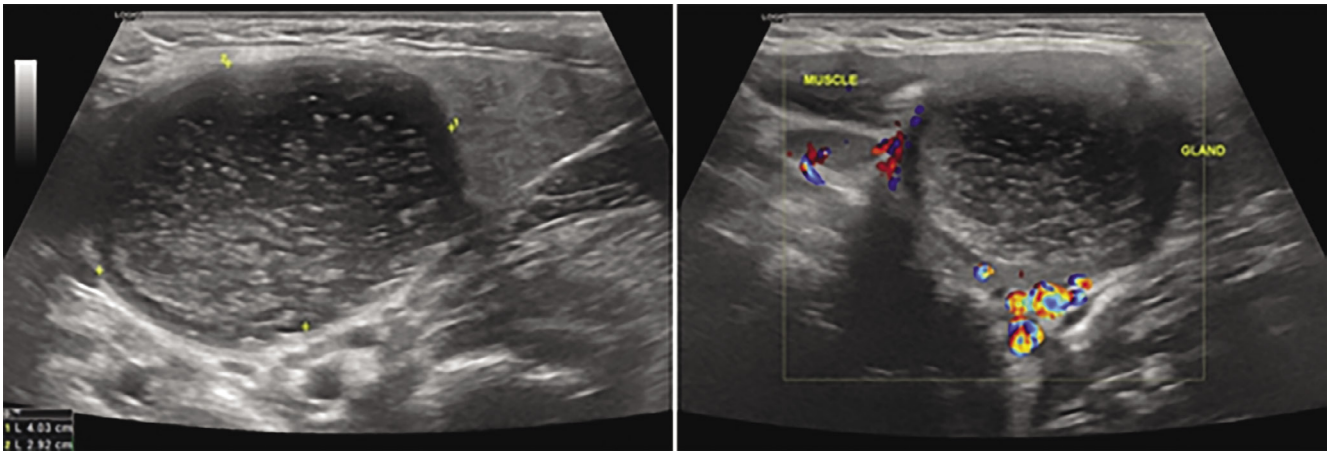


**Fig. 2.** *2nd cleft cyst: located in the submandibular region with thick mucous content of high echogenicity and without increased vascularity on color Doppler*

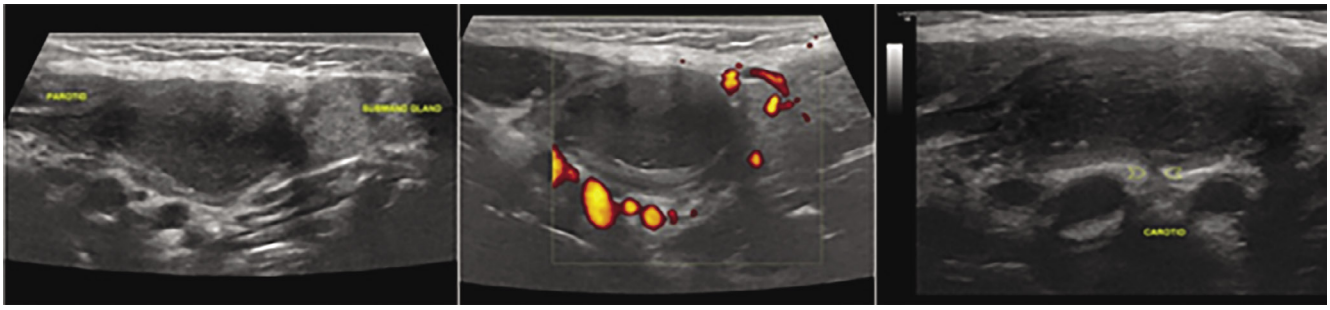


**Fig. 3.** *2nd cleft cyst: typical location of the lesion with anterior displacement of the submandibular gland, posterior displacement of the parotid gland, and inner pressure on the large vessels of the area. The presence of a small stem connecting the lesion to the carotid space is pathognomonic of the lesion*

result, the images were of very poor quality, and it was not easy for the observers to view and re-examine them for this study.

## Discussion

In most cases, BCC is identified clinically. The clinical differential diagnosis of BCC includes a large number of pathologies in the region, such as odontogenic infection, parotid swelling, tuberculous lymphadenitis, lipoma, cystic hygroma, carotid body tumors, thyroglossal duct cysts, suppurative lymphadenitis, branchial fistulas or sinus, dermoid cysts, neurofibroma, hemangioma, lymphangioma, teratoma, ectopic salivary tissue, pharyngeal diverticulum, laryngocele and plunging ranula[14–15]. Practically all pathologies that present with similar symptoms in the region should be included in the diagnosis[2–4]. Clinicians

must consider malignancies involving the lymph nodes, either primarily or secondarily, especially in elderly patients. Table 2 shows the differential diagnosis according to BCC types.

Besides the clinical symptoms and the appearance of the patients, various theories have been suggested in the literature regarding the origin of the BCC. These include incomplete obliteration of branchial mucosa, persistence of vestiges of the precervical sinus, thymopharyngeal ductal origin, and cystic lymph node origin. Many investigators have noted a link between the pharyngeal arches and branchial cysts[2]. The diagnosis of BCC is made initially by taking the patient's medical history and considering the clinical manifestations of the symptoms.

Pre-operative diagnostic procedures include CT, MRI, sonography, and fine-needle aspiration. In most cases, CT not only confirms the cystic nature of BCC but also determines the extent and anatomical relationship with adjacent structures.

MRI and CT scans are preferred in the evaluation of branchial cleft cysts, though with the advancements in ultrasonographic imaging and more recent features, the ultrasound should be considered as the initial examination of the lesions as a readily accessible and radiation-free technique.

The choice of the imaging technique also depends on regional preferences.

MRI is preferred over CT for first branchial cleft cysts and for parapharyngeal masses that may be second branchial cleft cysts. As a technique, it makes it easy to confirm with a high degree reliability the nature of the mass, and more precisely it defines the extent of the lesion and its relationship to the surrounding structures. Limitations of CT and MRI are that both are unable to distinguish a branchial cleft cyst from lymphangioma in children or metastatic squamous cell carcinoma from cervical nodes in adults.

Ultrasonography, despite being a commonly used imaging modality, adequately helps to evaluate the extent and depth of neck lesions, though it is not always as clear as the other imaging techniques. It is useful as a method where radiation exposure is to be avoided in patients, and in body locations where CT and MRI are not available.

More recently, Reynolds and Wolinski described uniform low- to medium-level echogenicity in cysts due to the presence of cholesterol crystals and cellular material. This feature was present in most of the cases analyzed and included in this study.

First branchial cleft cysts are less common and are anatomically related to the parotid gland. They present clinically just in front (of) or below the ear at the angle of the jawline[5,7,8]. The external sinus tract opening can be above the jawline (type I) or below the jawline in the upper neck above the level of the hyoid bone (type II). If there is an internal opening, it will be in the ear canal.

In the majority of first branchial arch cysts, cystic appearance and presentation are rarer when compared to fistulas and simple cavities.

Embryologically, the first branchial apparatus completes its development into the maxilla, mandible, external auditory canal, Eustachian tube, and portions of the middle ear structures by the sixth and seventh weeks of embryonic life. As the parotid gland and facial nerve develop later, a vestigial first branchial anomaly is located around them[8,16–18]. Thus, the first branchial cleft cyst can originate anywhere along the nasopharynx, middle ear, or external auditory canal, and it can extend down to the angle of the mandible. This specifies the location for the first branchial cleft cyst and facilitates the diagnosis.

Second branchial arch cysts are the most frequently occurring, as the fissure/cleft from which they develop (pharyngeal cleft) is the largest and the one that persists longer during embryonic development[2,5,19].

They are believed to be the most common type of branchial anomaly reported in the literature, accounting for 30 to 95% of all branchial apparatus anomalies. However, on reviewing the published literature that focused specifically on pediatric patients, we found that this frequently reported distribution may not in fact be accurately reflected in pediatric populations.

The second branchial apparatus forms the facial muscles, styloid process, and portions of the middle ear structure; the developmental process is complete by the sixth or seventh week of gestation. What is characteristic in embryology is that the second branchial cleft cyst, which is formed above the third arch, must pass between the internal and external carotid arteries. Then, the second branchial cleft cyst progressively moves upward and medially between the anterior margin of the sternocleidomastoid muscle and the tonsillar fossa. The location is the most frequent of all clefts in the area.

Bailey has classified second branchial cleft cysts (BCCs) into four types:[1]

- Type-I occurs anterior to the sternocleidomastoid muscle just deep to the platysma muscle;
- Type-II is the commonest type and occurs deep to the sternocleidomastoid and lateral to the carotid space;
- Type-III extends medially between the bifurcation of internal and external carotid arteries to the lateral pharyngeal wall; and
- Type-IV occurs in the pharyngeal mucosal space medial to the carotid sheath.

A second branchial arch cyst can occur anywhere in the lateral aspect of the neck but cysts of this type are most commonly seen at the floor of the mouth in the anteromedial border of the sternocleidomastoid muscle and the posterior margin of the submandibular gland[2–5]

On ultrasonographic examination, the lesion is usually located exteriorly and laterally to the internal jugular vein and caudally to the posterior belly of the digastric muscle.

It can be seen as a clear, round border lesion with liquid content and in most cases completely anechoic, with a thin well-defined wall around them.

Malformations of the third, and fourth branchial clefts are considerably less common, and they typically present clinically with recurrent and chronic neck inflammations[20–21]. They are both related to the pyriform sinus with those of the third cleft being above the superior laryngeal nerve and those of the fourth being below the nerve.

Fourth branchial cleft anomalies are generally sinus tracts or fistulae, and arise from the pyriform sinus, pierce the thyrohyoid membrane, and descend along the tracheoesophageal groove[21–23]. Most third branchial anomalies originate in the posterior compartment behind the sternocleidomastoid muscle. They may develop adjacent to or within the larynx. Upon imaging of the lesion, the diagnosis can be readily made, and no misdiagnosis to other similar symptoms and clinical pathologies is to be expected[23]. Radiographic imaging with the use of ultrasonography is clear and definitive of the diagnosis in BCCs. However, physicians should be aware of the condition and always include it in their diagnosis, when relevant.

## Conclusions

The etiology of BCC is has not been fully elucidated, through the majority of authors and opinions suggest that they originate from the branchial apparatus or the lymphoid tissues. The precise embryological origin can be predicted in most cases. However, because of similar clinical appearance the differential diagnosis from other lesions of the region should be carefully done, with ultrasound imaging recommended as a diagnostic modality.

### Conflict of interest

*The authors do not report any financial or personal connections with other persons or organizations which might negatively affect the contents of this publication and/or claim authorship rights to this publication.*

### Author contributions

*Original concept of study: AD, KO. Writing of manuscript: AD, KO. Analysis and interpretation of data: AD, MO, KO. Final acceptation of manuscript: AD, MO, KO. Collection, recording and/or compilation of data: AD, MO, KO. Critical review of manuscript: AD, KO.*

## References


1. Bailey H: Branchial Cysts and Other Essays on Surgical Subjects in the Facio-Cervical Region. H.K. Lewis & Company, London 1929.
2. Work WP: Newer concepts of first branchial cleft defects. Laryngoscope 1972; 82: 1581–1593.
3. Acierno SP, Waldhausen JHT: Congenital cervical cysts, sinuses, and fistulae. Otolaryngol Clin North Am 2007; 40: 161–176, vii–viii.
4. Choi SS, Zalzal GH: Branchial anomalies: a review of 52 cases. Laryngoscope 1995; 105: 909–913.
5. Ford GR, Balakrishnan A, Evans JN, Bailey CM: Branchial cleft and pouch anomalies, J Laryngol Otol 1992; 106: 137–143.
6. Bajaj Y, Ifeacho S, Tweedie D, Jephson CG, Albert DM, Cochrane LA *et al.*: Branchial anomalies in children. Int J Pediatr Otorhinolaryngol 2011; 75: 1020–1023.
7. Chandler JR, Mitchell B: Branchial cleft cysts, sinuses, and fistulas. Otolaryngol Clin North Am 1981; 14: 175–186.
8. Frazer JE: The nomenclature of diseased states caused by certain vestigial structures in the neck. Br J Surg 1923; 11: 131–136.
9. Schroeder JW Jr, Mohyuddin N, Maddalozzo J: Branchial anomalies in the pediatric population. Otolaryngol Head Neck Surg 2007; 137: 289–295.
10. Thomaidis V, Seretis K, Tamiolakis D, Papadopoulos N, Tsamis I: Branchial cysts. A report of 4 cases. Acta Dermatovenerol Alp Panonica Adriat 2006; 15: 85–89.
11. Mandell DL: Head and neck anomalies related to the branchial apparatus, Otolaryngol Clin North Am 2000; 33: 1309–1332.
12. Nicollas R, Guelfucci B, Roman S, Triglia JM: Congenital cysts and fistulas of the neck. Int J Pediatr Otorhinolaryngol 2000; 55: 117–124.
13. Hsieh Y-Y, Hsueh S, Hsueh C, Lin J-N, Luo C-C, Lai J-Y *et al.*: Pathological analysis of congenital cervical cysts in children: 20 years of experience at Chang Gung Memorial Hospital. Chang Gung Med J 2003; 26: 107–113.
14. D'Souza AR, Uppal HS, De R, Zeitoun H: Updating concepts of first branchial cleft defects: a literature review. Int J Pediatr Otorhinolaryngol 2002; 62: 103–109.
15. Coppens F, Peene P, Lemahieu SF: Diagnosis and differential diagnosis of branchial cleft cysts by CT scan, J Belge Radiol 1990; 73: 189–196.
16. Chen M-F, Ueng S-H, Jung S-M, Chen Y-L, Chang K-P: A type ii first branchial cleft cyst masquerading as an infected parotid Warthin's tumor. Chang Gung Med J 2006; 29: 435–439.
17. Hickey SA, Scott GA, Traub P: Defects of the first branchial cleft. J Laryngol Otol 1994; 108: 240–243.
18. Triglia J, Nicollas R, Ducroz V, Koltai PJ, Garabedian EN: First branchial cleft anomalies: a study of 39 cases and a review of the literature. Arch Otolaryngol Head Neck Surg 19998; 124: 291–295.
19. Papay FA, Kalucis C, Eliachar I, Tucker HM: Nasopharyngeal presentation of second branchial cleft cyst. Otolaryngol Head Neck Surg 1994; 110: 232–234.
20. Nicoucar K, Giger R, Jaecklin T, Pope HG Jr, Dulguerov P: Management of congenital third branchial arch anomalies: a systematic review. Otolaryngol Head Neck Surg 2010; 142: 21–28.e2.
21. Pereira KD, Losh GG, Oliver D, Poole MD: Management of anomalies of the third and fourth branchial pouches. Int J Pediatr Otorhinolaryngol 2004; 68: 43–50.
22. Nicoucar K, Giger R, Pope HG Jr, Jaecklin T, Dulguerov P: Management of congenital fourth branchial arch anomalies: a review and analysis of published cases. J Pediatr Surg 2009; 44: 1432–1439.
23. Godin MS, Kearns DB, Pransky SM, Seid AB, Wilson DB: Fourth branchial pouch sinus: principles of diagnosis and management. Laryngoscope 1990; 100: 174–178.