

Original Article

Multidisciplinary Management of Third and Fourth Branchial Remnants at a Pediatric Hospital

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ABSTRACT

Background: Third and fourth branchial remnants account for less than 4% of all branchial anomalies. Despite the availability of diverse imaging techniques, underdiagnosis is frequent. The objective of this study is to describe the experience of the multidisciplinary treatment of patients with third and fourth branchial remnants in a tertiary care pediatric hospital in Colombia. This is a retrospective descriptive observational case series study. The objective was to describe diagnostic and therapeutic variables of children with third and fourth branchial remnants treated at the Fundación HOMI from January 2018 to January 2023.

Results: Eight patients met the inclusion criteria. The female to male ratio was 3:1. The mean age at diagnosis was 75.1 months (22-177, SD 57.3). All lesions were located on the left neck. All patients presented with an abscess at the time of diagnosis. The initial treatment in 38% of patients was based on a mixed approach involving endoscopic cauterization of the fistula and surgical excision of the tract under ultrasound guidance. An average of 1.6 cauterizations were performed in each case (0-3, SD 1). The mean duration of follow-up was 108.6 days (0-556, SD 192.2), and at the end of this period, 75% of the patients were asymptomatic.

Conclusions: The diagnosis of pyriform sinus fistulas warrants a high level of suspicion. A combined approach of endoscopic strategies and ultrasound-guided surgery increases the likelihood of success and may reduce morbidity.

Keywords: Branchial arch; interventional ultrasound; pyriform sinus; suppurative thyroiditis, trichloroacetic acid.

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BACKGROUND

Third and fourth branchial remnants account for less than 4% of all branchial anomalies diagnosed in children^[1,2,3]. Their clinical presentation depends on the age of the patient at the time of diagnosis. In newborns, it is common to see a

mass with a compressive effect on the upper airway^[4], while older children usually present with cervical abscesses and suppurative thyroiditis^[1,2].

The pathophysiology of these lesions involves a failure in

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the regression of the transient fetal structures that contribute to the development of the definitive anatomy of the head and neck. The branchial apparatus develops between the fourth and seventh week of gestation, which involves elements of the endoderm, mesoderm, and ectoderm^[5]. These structures' partial or complete persistence can lead to the appearance of cysts, sinuses, or cervical fistulas. Third and fourth branchial remnants are located in the tract between the pyriform fossa in the hypopharynx and the superior pole of the left thyroid lobe in more than 95% of the cases^[5,6].

The anatomical structure typically described to identify the trajectory of pyriform sinus fistulae (PSF) is the recurrent laryngeal nerve^[5]. However, cervical-mediastinal trajectories surrounding the great vessels of the thoracic operculum are rarely successfully depicted during clinical assessment^[1,5]. A possible explanation is that these lesions are remnants of the thymopharyngeal duct, which originates in the pyriform fossa and descends to the superior mediastinum, in proximity to the ipsilateral thyroid lobe, which may account for the high incidence of infections in the thyroid gland^[7].

Despite the multiple diagnostic modalities available to evaluate neck masses in children, PSFs are frequently underdiagnosed, so a high level of suspicion is necessary in these cases. Proper diagnostic and therapeutic approach can reduce the likelihood of recurrence of these lesions^[8,9]. Therefore, the objective of this study is to describe the experience of the multidisciplinary treatment of patients with third and fourth branchial remnants in a tertiary care pediatric hospital in Colombia.

METHODS

This is a retrospective descriptive observational case series study of pediatric patients with third and fourth branchial remnants was conducted. The institutional ethics committee endorsed the study by means of Minutes No. 71 550-23. The databases of the Pediatric Surgery and the Ear, Nose, and Throat (ENT) departments at Fundación HOMI in Bogotá, Colombia, were accessed, and a retrospective review of medical records was carried out.

Patients under 18 years of age who had a pyriform sinus fistula detected on endoscopic examination in the period from January 01, 2018, to January 31, 2023, were included. Cases involving patients with branchial remnants that were not in the third or fourth arches were excluded.

At our institution, patients with cervical abscess are assessed with a neck ultrasound. Sometimes, during the acute infection, CT scan is performed to assess the extent of the infection and its relation with adjacent organs. Once the inflammatory process is under control, an MRI is made in order to clarify the anatomy of the remnant. Every patient with history of left cervical abscess near the thyroid gland had an endoscopic evaluation due to FSP suspicion; patients with demonstrated fistula (included in our study) underwent a combined (open and endoscopic) approach. Chemical

cauterization was performed with trichloroacetic acid (50% concentration - Fagron®), applied to the proximal fistula tract with a cottonoid after brushing the mucosa endoscopically. During open approach, methylene blue was injected to the cyst in order to identify the complete fistula tract and avoid missing any of its segments during surgical resection.

A database was prepared after collecting the variables (demographic, clinical and surgical) described for each case. No statistical analysis was performed due to the study design; however, descriptive measures for quantitative data (mean, range, and median) and qualitative data (frequencies expressed as percentages) were calculated.

RESULTS

A total of 8 patients met the inclusion criteria during the study period, of whom 75% were female. The mean age of the patients at diagnosis was 75.1 months (22-177, SD 57.3). The mean time of progression at the moment of diagnosis was 26.3 months (2-110, SD 34.9). All lesions were located on the left neck.

The first manifestation in all patients was a mass with inflammatory changes, meaning that 100% had at least one abscess before surgery. On average, 5.1 abscesses per patient (2-18, SD 5.3) were reported from the initial diagnosis until the last assessment. The preoperative evaluation was conducted using ultrasound in 87.5% of cases, CT scans in 50%, and magnetic resonance imaging (MRI) in 62.5%. In all cases, the images allowed to confirm the presence of an abscess in close proximity to the left lobe of the thyroid gland. Moreover, contrast studies were useful for characterizing fistulous tracts towards the pharynx in some cases. Only one of the cases underwent an upper GI series with a water-soluble contrast medium, in which an esophagocutaneous fistula was identified in a patient with a history of multiple abscesses and recurrences despite endoscopic and surgical treatment.

The initial treatment provided to 3 (38%) of the patients involved a mixed approach with endoscopic cauterization of the fistula and ultrasound-guided surgical excision of the cyst. In comparison, 37.5% were exclusively treated with endoscopy and 25% by open approach only. Chemocauterization was performed under general anesthesia, with suspension laryngoscopy, making a thorough exploration of the hypopharynx and bilateral pyriform fossa. In the case of redundant tissue, the apex of the pyriform sinus was explored with an endoscopic probe until the fistulous orifice was located. After confirming the presence of the fistula, brushing was carried out to create mucosal abrasion, and 50% trichloroacetic acid (TCA) was applied to the exposed tissue to effectively cauterize the edges of the opening (Figure 1).

Following the endoscopic procedure, in patients with a mixed approach, an ultrasound scan of the left neck was performed using a high frequency hockey-stick linear transducer along the axial and longitudinal axes. Exploration started at the scar from the previous surgical drains and

extended across the front edge of the sternocleidomastoid muscle towards the head. Among the observations, a cystic lesion displaying a hyperechoic contour and containing some mobile echoes within it (n=5), or a diffuse, ill-defined, hyperechoic lesion along the fistula trajectory (n=3), were

identified. In certain cases, intraoperative ultrasound was employed to assess the distance from the skin's surface to the lesion and to ascertain the extent of resection when dealing with a severe scarring process.

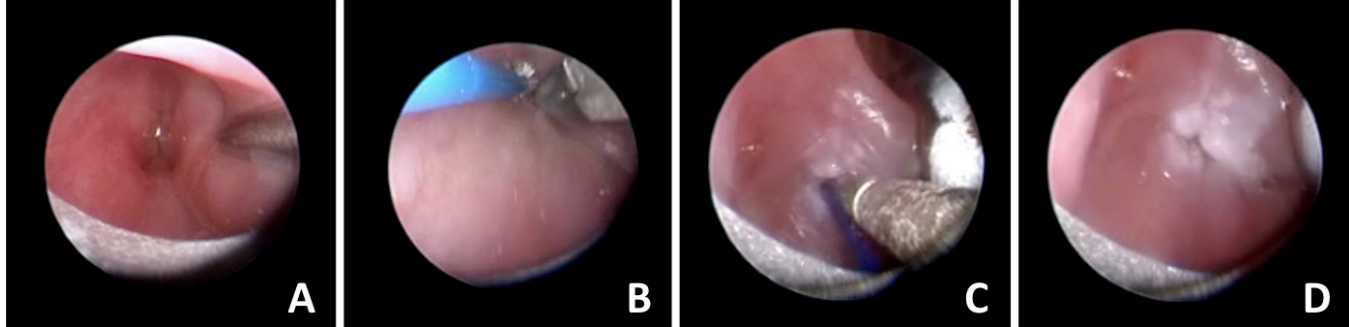


Fig. 1: Topical application of 50% trichloroacetic acid. A) Location of the internal orifice of the fistula in the left pyriform sinus. B) Brushing of the tract to expose the mucosa for sclerosing agent application. C) Application of 50% TCA to the prepared mucosa. D) Confirmation of cauterization of the orifice edges, resulting in the desired outcome.

In 87.5% of the cases, an endoscopic follow-up was performed after the first procedure, showing persistence of the fistula in 6 out of 7 cases. The incidence of complications was 50%, including recurrent infections (n=1, 12.5%), recurrent cyst (n=2, 25%), and surgical site infection (n=1, 12.5%). An average of 1.6 cauterizations were performed in each case (0-3, SD 1). The mean follow-up time after the last intervention was 108.6 days (0-556, SD 192.2), at which time 75% of the patients were asymptomatic. Patients in whom recurrence was documented continue to be followed up on an outpatient basis, with plans for new endoscopic exploration and, if necessary, cervical reintervention.

DISCUSSION

PSFs are a rare entity. The present study found that 75% of these cases occurred in females and that the age range at diagnosis was between 1.8 and 14.8 years, which coincides with the literature;^[1,9] contrary to some reports,^[4,10] no case was reported in neonates. All lesions were located in the left neck, which is similar to previous reports describing nearly 90% of left-sided lesions^[3,9,10] (Figure 2). Other studies have documented the presence of abscesses at the time of diagnosis in 50% to 75% of cases^[4,8,11]. In this series, all cases had multiple cervical abscesses prior to definitive diagnosis (Table 1), there were no cases of salivary fistula amongst our patients

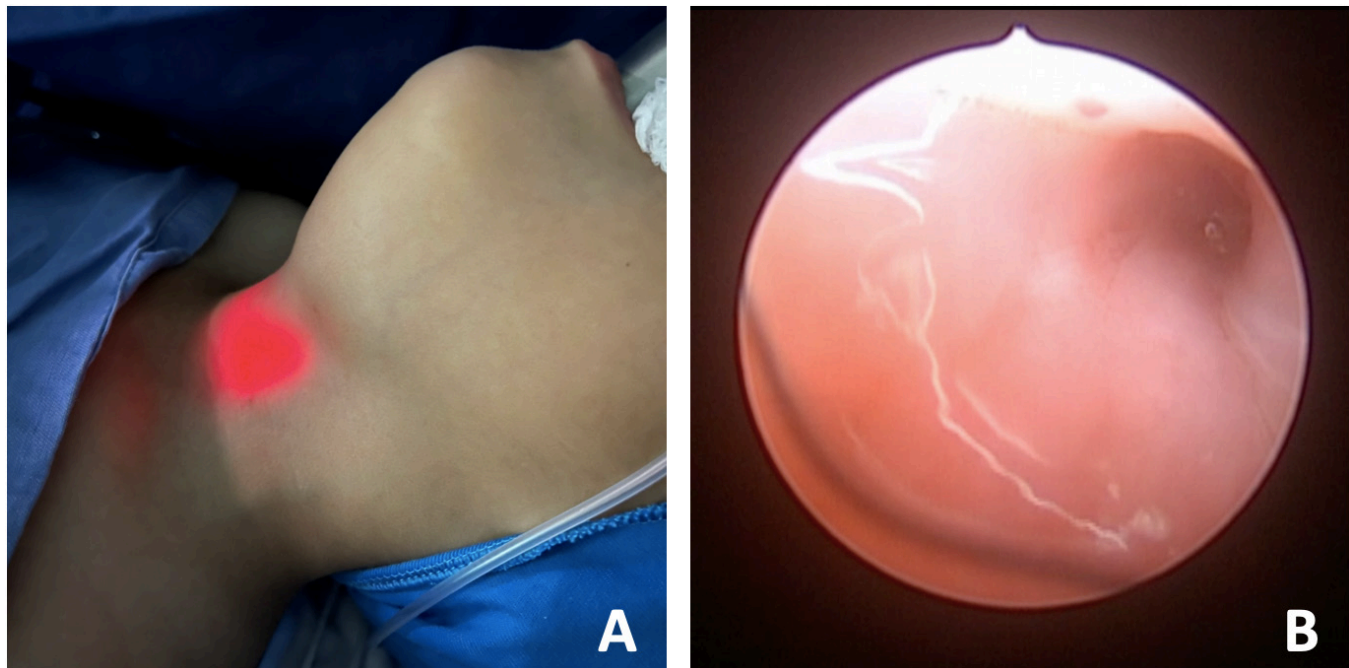


Fig. 2: A) External localization of a left pyriform sinus fistula cannulated with endoscopic examination lens. B) Fistulous orifice identified in suspension laryngoscopy.

Table 1: Clinical characterization of patients with pyriform sinus fistula

Case	Age at diagnosis (years)	Sex	Laterality	Time to diagnosis (months)	Initial presentation	Number of abscesses from diagnosis to last follow-up
1	2	F	Left	2	Abscess	3
2	7	F	Left	5	Abscess	3
3	14	F	Left	12	Abscess	3
4	3	M	Left	14	Abscess	18
5	4	M	Left	17	Abscess	5
6	2	F	Left	22	Abscess	2
7	3	F	Left	28	Abscess	2
8	11	F	Left	110	Abscess	5

The most widely used diagnostic modality for the initial assessment of these patients was ultrasound, considering that it is the most available, least expensive tool, with no ionizing radiation exposure, and very useful for characterizing inflammatory lesions of the neck, as described in previous publications^[4,8]. However, ultrasound rarely allows establishing a definitive diagnosis^[4,6] and, as a result, other imaging modalities may be indicated depending on the time of disease progression. CT scan gains importance in the scenario

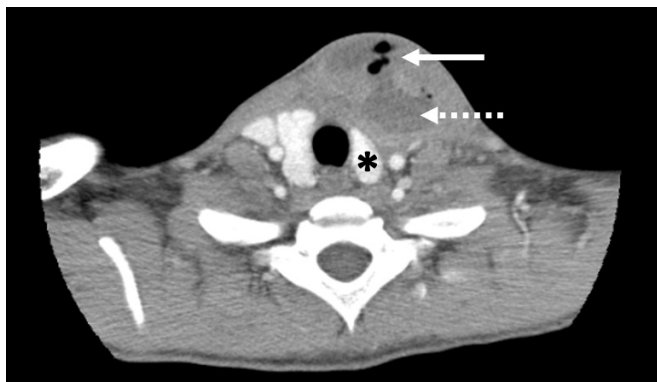


Fig. 3: Overinfected fourth branchial remnant with large cervical abscess in CT scan. Gas content within the fluid collection (solid arrow). Liquid content within the fluid collection (dashed arrow) in proximity to the left thyroid lobe (asterisk).

Some authors recommend the use of esophagogram with water-soluble contrast medium, alleging an adequate diagnostic yield^[2,4,6,11]. However, given the high incidence of infection and re-infection in the patients assessed in this series, gastrointestinal tract x-ray was not the modality of choice for initial work-up due to the likelihood of obliteration of the fistulous tract secondary to acute inflammatory changes, which may result in false negatives. Esophagogram was performed in only one case, in which recurrence was documented despite the use of various therapeutic strategies, resulting in the finding of a pharyngocutaneous fistula after multiple interventions.

Li *et al.*^[2] designed a classification system to group lesions based on their morphology. Type I lesions include cysts with or without external sinus; type II lesions are those with an initial internal sinus; type III lesions involve an internal sinus with thyroid abscess; and type IV lesions are

of acute infection to assess the extent of the lesion and its relationship with other structures of the neck^[4,6] (Figure 3); in the present study, it was performed in 50% of the patients. On the other hand, contrasted MRI may have a higher diagnostic yield in the infection-free setting, when there is a greater likelihood of identifying the fistulous tract between the hypopharynx and the thyroid gland^[8,12]; in this study, it was performed in 62.5% (n=5) of the cases, identifying the fistula in 4 of them (Figure 4).

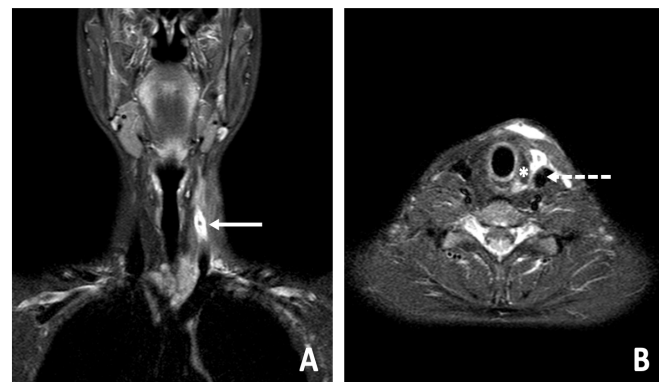


Fig. 4: A) Cervical MRI, T2 sequence showing hyperintense fistulous tract with air content (solid arrow). B) Same image in axial plane projection. Anatomical relations of the fistula in proximity to the left thyroid lobe (asterisk) and the carotid sheath (dashed arrow).

refractory fistulas with multiple recurrences. Since branchial remnants from different origins may be mistaken for each other, only those with endoscopically demonstrated fistulae were included in the present study. Given that Fundación HOMI is a referral hospital, all patients were treated after they had already developed infectious complications, so all our patients had type III and IV lesions, explaining the high rate of complications observed, which has also been reported in the literature^[13,14].

Over the last five years, our hospital's approach has been consolidated through the joint assessment of each case by an ENT specialist and a pediatric surgeon in order to improve endoscopic evaluation and the accuracy of open surgery. Whenever it has been possible to use the mixed approach as the initial treatment, procedures are performed simultaneously by both specialists to discuss the findings and technical details.

There are multiple alternatives for endoscopic cauterization of the fistulous orifice. Several authors have stated their preference for laser devices^[10,15,16,17], although multiple strategies have been proposed to achieve definitive obliteration of the tract, including the application of fibrin glue and sclerosant agents, or the use of endoscopic sutures^[1,15,17,18]. In the present study, 50% TCA was used due to the overall experience of the staff in the treatment of other conditions such as recurrent tracheoesophageal fistula. Success rates with the various agents vary depending on the series evaluated; however, due to the low prevalence of the disease, the available evidence is not statistically significant to establish the superiority of one over the other, therefore there is no standard treatment for this condition.

Regarding open surgery, the approach used involved transverse cervical incision and resection of the scar from previous surgical drainage. The location of the lesion was assessed with intraoperative ultrasound, which is a useful tool to guide dissection in cases of dense fibrosis and chronic scarring processes. High ligation of the tract near the origin of the pyriform fossa was performed near the inferior thyroid horn, but identification of the anatomic landmarks was limited by chronic inflammation. Contrary to what has been described in the literature^[1,11,19], hemithyroidectomy was not routinely performed in patients with inflammatory involvement of the gland.

Most of the patients included in this study had no symptoms during the last assessment, with an average of 1.6 endoscopic interventions with or without open surgery for each case. The complication rate was high, but complex cases with late diagnosis and up to 18 episodes of abscesses before definitive treatment were included. A mixed approach executed by a multidisciplinary group may be successful if it achieves to optimize diagnostic opportunity, expedite referral to the center of expertise, and minimize the time elapsed from the first infection to surgery.

One of the limitations of the present study is the sample size, which made it impossible to perform a statistical analysis that would allow us to generalize conclusions regarding the diagnosis and treatment of this condition. Furthermore, there is a risk of selection bias with respect to the inclusion criteria considered in the study, considering that an acute inflammatory process may alter the findings of the endoscopic assessment of the pyriform fossa, making the presence of a fistulous orifice imperceptible as a result of local inflammation. All the foregoing may have contributed to the non-inclusion of patients with third and fourth branchial remnants in whom the presence of PSF could not be documented.

CONCLUSIONS

Third and fourth branchial remnants are associated with high morbidity due to frequent relapses as a consequence of incomplete obliteration of the lesion achieved with isolated therapeutic modalities. The implementation of a multidisciplinary approach which involves complete surgical

resection of both the cyst and the fistula tract with ultrasound guidance and endoscopic cauterization of the fistula.

DECLARATIONS

Ethics approval and consent to participate: The institutional ethics committee endorsed the study by means of Minutes No. 71 550-23. Due to the design of the study, no informed consent was required.

CONSENT FOR PUBLICATION

Not applicable.

AVAILABILITY OF DATA AND MATERIAL

All data generated or analyzed during this study are included in this published article.

COMPETING INTERESTS

The authors declare that they have no competing interests.

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AUTHORS' CONTRIBUTIONS

All authors participated in study conception and protocol elaboration. Data collection was carried out by M.E.R.B. Manuscript drafting was carried out by L.C.G.V. Figure preparation was carried out by A.I.L.L and J.J.V.H. All authors read and approved the final manuscript.

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Not applicable.

ABBREVIATIONS

(PSF): Pyriform sinus fistula, **(TCA):** Trichloroacetic acid, **(CT):** Computed tomography, **(MR):** Magnetic resonance, **(POCUS):** Point of care ultrasound.

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