Neonatal vs Delayed-Onset Fourth Branchial Pouch Anomalies

Therapeutic Implications

Nicolas Leboulanger, MD; Katell Ruellan, MD; Jérôme Nevoux, MD; Stéphane Pezzettigotta, MD; Françoise Denoyelle, MD, PhD; Gilles Roger, MD; Érée-Noël Garabedian, MD

Objectives: To determine the presentation of third or fourth branchial pouch anomalies in various age groups of children and evaluate endoscopic cauterization as a treatment technique.

Design: Retrospective study of patients treated from 2000 to 2009.

Setting: Tertiary care children’s hospital.

Patients: Pediatric patients aged 0 to 18 years (mean age, 5.5 years), including 5 neonates.

Interventions: Endoscopic and/or open surgical management of third and fourth branchial pouch anomalies; clinical and endoscopic follow-up.

Main Outcome Measures: Absence of clinical recurrence; closure of the sinus tract.

Results: Two forms of presentation were identified: a neonatal form, characterized by a voluminous and compressive cervical mass (5 of 20 [25%]) and a childhood form, presenting as a cervical abscess (15 of 20 [75%]). The vast majority of our patients regardless of presentation were treated endoscopically (n=19), with a success rate of 68% (13 of 19) after 1 procedure, 79% (15 of 19) after 2 procedures, and 89% (17 of 19) after 3 procedures. Neonatal and adult presentations require slightly different therapeutic approaches.

Conclusions: Third and fourth branchial pouch anomalies can present in 2 distinct forms: a neonatal form and a childhood form. The endoscopic technique should be the favored approach for both forms: whenever possible, in view of its simplicity, rapidity, and the lack of serious postoperative complications. Recurrences can be treated by repeated cauterization using the same technique, with good long-term outcomes. An age-based management algorithm has been developed.

B RANCHIAL ARCH ABNORMALITIES represent approximately 20% of cervical masses in children.1 Such lesions developing from the third and fourth branchial pouches are less frequent (3% to 10%).1,2 The distinction between third and fourth branchial pouch abnormalities can only be established at the time of dissection because their clinical presentations are similar. From a practical point of view, the anatomic differences between these 2 lesions are of little significance because their clinical presentations and management strategies are often the same. From 90% to 100% of these sinus tracts are situated on the left side of the neck.3-5 In older children, they often present as abscesses (Figure 1), cervical masses, or rapidly relapsing thyroiditis, usually situated on the left. In the neonatal period, the presentation is usually as a cystic mass or an abscess, which may lead to dyspnea with stridor, dysphagia, and feeding difficulties.6-8

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The diagnostic workup of these lesions includes imaging. Ultrasonography can be used to identify the cervical collection, but to show the path of the tract either computed tomography (CT) or magnetic resonance imaging (MRI) is required5,9 (Figure 2). The presence of an air bubble in the lesion is also characteristic of this malformation and is more often seen in the neonatal form.10 The best diagnostic examination is endoscopy, which allows confirmation of the diagnosis by direct visualization of the orifice of the tract in the piriform fossa and the Betz fold (Figure 3).2,11,12 The classic management described in the literature is surgical resec-
tion using a cervical approach. However, in the last 15 years a less invasive treatment has evolved, namely, endoscopic cauterization limited to the sinus tract orifice.

METHODS

We performed a retrospective study looking at the medical records of 20 patients treated in our department for a sinus tract of the fourth branchial pouch from January 2000 to May 2009. This study received the approval of the institutional review board of our hospital.

RESULTS

The 15 girls and 5 boys included in the study all had a unilateral form of the condition. Age at diagnosis varied from day of birth (5 neonatal cases) to 18 years; the mean age was 5.5 years, and the median age was 4 years. The lesion was left sided in 18 of the 20 cases (90%). The clinical and treatment characteristics are summarized in the Table.

The clinical presentation in 4 of the neonates was a cervical mass compressing the larynx without evidence of infection. All other patients (n=16) had an infected or inflammatory lesion. The interval between the first clinical signs and/or symptoms and the diagnosis ranged from 2 days to 4 months (mean interval, 21 days). All the patients with an abscess underwent radiologic imaging—in total, 85% of the patients underwent a CT scan (17 of 20). Endoscopy established the diagnosis in all cases.

All patients with an infected lesion (n=12 [60%]) were treated with a 10-day course of antibiotics (amoxicillin plus clavulanic acid), which was followed by either immediate cauterization of the sinus tract orifice (9 of 12 [75%]) or drainage (3 of 12 [25%]), which then allowed subsequent endoscopic cauterization of the sinus tract orifice. In 19 cases the treatment was endoscopic (95%), with a success rate of 68% (13 of 19) following 1 intervention; 79% (15 of 19) after 2 interventions; and 89% (17 of 19) after 3 procedures. No patient had more than 3 endoscopic cauterizations.

In 1 neonatal case (patient 4), the initial treatment was open surgery owing to a cervical mass causing clinically significant symptoms. Another neonate (patient 2) had an initial endoscopic intervention but later underwent a cervicotomy owing to an early recurrence of the cervical mass on postoperative day 3.

A nasogastric tube was left in situ in 3 cases for 2 to 8 days (mean, 4.3 days). The presence or absence of a nasogastric tube did not appear to affect postoperative complications or later recurrences.

A follow-up endoscopy was systematically performed in all cases treated by endoscopy, with an interval of 8 days to 5 months (mean interval, 5.7 weeks). The sinus tract relapsed in 7 cases (37%) within a mean interval of 6 weeks (time range to relapse, 3 days to 3 months). In 3 patients, the relapse was diagnosed following formation of an abscess that appeared 2 to 3 months after treatment, although the follow-up endoscopies in all 3 cases had shown only an area of scarred mucosa and the closure of the tract. Three patients were diagnosed with an “anatomical” relapse (persistence of the sinus tract orifice without clinical symptoms) at the follow-up endoscopy (at 1 and 2 months after surgery). Patient 2 had an early recurrence of the cervical mass, leading to open surgery.
At last follow-up none of the 7 patients who experienced a relapse showed any new symptoms. Findings of follow-up endoscopies were normal for all patients treated via a cervical approach (mean follow-up time, 4.1 months). After 2 further endoscopic interventions, the overall failure rate for endoscopic treatment was 11% (2 of 19). None of the 4 patients treated with a cervicalotomy experienced relapse. Average follow-up was 23 months (range, 6 months to 5 years).

To our knowledge, the present series of patients treated for a sinus tract of the fourth branchial pouch is one of the largest series in the literature (20 cases between 2000 and 2009). It is also the largest series to our knowledge in which most patients were treated with an endoscopic approach (19 of 20 cases [95%]). There was also a large number of neonatal cases (5 of 20 cases [25%]) in our series, compared with 0% to 20% in other studies.1−3,4,11−13

Our population showed a marked female bias, with a ratio of 3 girls for each boy. The literature is not clear on the issue of incidence with respect to sex.1−3,4,12−15

In 18 of 20 cases, the lesion was located on the left side (90%), and in 13 of 18 cases, the clinical presentation was principally as cervical inflammation or infection (72%). These results are similar to those reported in the literature. The mean age for the development of the first clinical signs was 5.2 years, and this varies between 4 and 7 years in the literature.1−4,11−16

The imaging technique of choice for fourth branchial pouch anomaly is usually CT because CT is cheaper than MRI, generally more readily and widely available, and only rarely requires a general anesthetic. However, CT exposes the patient to irradiation and thus cannot be repeated at will. The tract can be visualized by using contrast material,1−3,5,13,17 but while this test is highly specific, its sensitivity is only between 50% and 80%. This poor sensitivity can be attributed to periorificial edema, which leads to obstruction of the sinus during acute infection and so prevents passage of the contrast material.3,17

Our patients underwent a rapid diagnostic workup with a short mean interval from clinical presentation to diagnosis (21 days). This is substantially shorter than the intervals described in the literature, in which cases many patients had several infective episodes before the correct diagnosis was considered. For example, in the series reported by Rea et al,11 the interval between the first clinical signs and the diagnosis was 6 years on average, and a mean of 6 interventions were performed per patient before the final diagnosis was made. In the series described by Shrime et al,3 4 of 6 patients had recurrent abscesses before the diagnosis was made. Similarly, James et al1 reported that 14 of 17 patients had at least 2 surgical procedures and 6 of 17 had recurrent infections. Other series report several cases of recurrent cervical infections before the final diagnosis was established.2,4,10,12,15

Analysis of the results of the present study and those reported in the literature allows identification of 2 main clinical forms of this condition representing different clinical manifestations: a neonatal and a childhood form. The neonatal form manifests as a very large cervical mass that may or may not be infected and readily compresses surrounding structures. There is often a bubble of air visible within the mass on imaging. The mass may cause dyspnea with stridor and feeding difficulties.3,4,6−8,18 In our experience, histologic examination of the resected specimen shows the presence of malpighian epithelium lining the tract along its path as well as the pouch, as described in the literature.6,7,14 This contrasts with the histologic findings of cases presenting later in childhood in which such epithelium lining is restricted to the tract itself.12,14

<table>
<thead>
<tr>
<th>Patient No./Age</th>
<th>Clinical Feature</th>
<th>Initial Approach</th>
<th>Endoscopic Procedure</th>
<th>Recurrence</th>
<th>Secondary Approach</th>
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<tbody>
<tr>
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<tr>
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<tr>
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<td>CO2 laser</td>
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<tr>
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<td>Endoscopic</td>
<td>Thulium laser</td>
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</table>

Abbreviations: CO2, carbon dioxide; NA, not applicable; NB, newborn.
Five of our patients were in this neonatal category. In one case, endoscopic management proved to be inadequate, and resection via a cervicotomy was necessary. In another case, early recurrence of the cervical mass after needle aspiration led to immediate open surgery without an initial endoscopic approach. The high rate of failure of endoscopic treatment in this scenario is noteworthy (40% [2 of 5]). We believe that surgical dissection of the cyst and the tract was easier in these 2 cases. The literature reports treatment of this form of the condition via cervicotomy, which allows a straightforward dissection of the cervical mass all the way to the bottom of the piriform fossa. The associated complications are minor: dislocation of the arytenoid during endoscopy and transient paralysis of the larynx. To our knowledge, there is no published report of the neonatal condition being treated endoscopically.

The second form of the disease presents in later childhood, is smaller than the neonatal form, and presents with a cervical abscess associated with local clinical features. In general, an air bubble is not present on imaging. In our series, the initial treatment of this form was endoscopic excision of the lesion via cervicotomy. The most frequent complication described in the literature is a transient or permanent paralysis of the larynx, reported in 0% to 33% of cases. This occurs more often when the tract of the sinus to be dissected is encased in scar or inflammatory tissue. Recurrence is limited to cases in which complete excision of the tract cannot be performed, which occurs most often when severe trauma is undertaken at the time of active inflammation and the tract is difficult to identify. The recurrence rate varies from 0% to 16% during a follow-up range from 2 months to 4 years.

Toward the end of the 1990s, a new surgical endoscopic technique was developed involving cauterization of the sinus tract orifice at the base of the piriform fossa using local coagulation, thereby destroying the tract, avoiding a subsequent adjacent “cyst” and theoretically leading to its involution. While intuitively this approach might seem inadequate, it has been used to achieve real therapeutic successes without systematic relapses or abscess formation. In general, cases of short epithelial sinus tracts are associated with infection in the surrounding tissue without an actual abscess wall. However, endoscopic management of a malformation consisting of a sinus tract connecting with a cervical cystic cavity with its own epithelial wall should, logically, lead to failure (eg, cases of massive cervical masses in neonates).

A number of treatment techniques have been reported in the literature. Verret et al have described the introduction of a balloon catheter after dilation of the sinus tract orifice to allow endoscopic cautery. Electrocoagulation was also used by Jordan et al with the aid of a diathermy probe. Alternatively, Sayadi et al used a low-power diode laser, while 2 other groups used chemical cautery with trichloroacetic acid. Kim et al and Pereira and Smith performed chemical cautery by introducing a stick of silver nitrate for 3 to 4 seconds into the sinus tract and at the sinus tract orifice. Finally, there is one report of the use of fibrin glue.

In our series, electrocauterization was performed with a diathermy point in 2 cases and by laser in 17 cases (carbon dioxide laser in 13 cases and a thulium laser in 4—thulium laser technology has only recently become available). A uniform approach was always taken: laryngoscopy was performed under general anesthesia, and orotracheal intubation was used to allow assisted ventilation. The mucosal fold of Betz was exposed and cut (with laser or cold instruments), thereby providing a large field of view of the path of the sinus tract following exposure of its medial aspect. Next, the tract was thoroughly coagulated (by aiming beam for the carbon dioxide laser and fiber for the thulium laser), including deep within the piriform fossa (Figure 4). The usual laser setting was 3 W in superpulse mode. Sayadi et al mention the presence of the fold of Betz but do not comment on sectioning it. However, the surgical sectioning of this structure is important because sectioning facilitates the field of view of the sinus tract and hence allows a more definitive cauterization. No complications, other than recurrence, were reported either in our series or in cases reported in the literature.

Our preferred approach is laser because of its ease of use, accuracy, and low level of thermal diffusion. In our opinion, coagulation by diathermy is too imprecise and carries the risk of damaging the inferior laryngeal nerve
by thermal diffusion. However, this has yet to be confirmed by further studies.

In the literature reports, the recurrence rate with this technique varies from 0% to 25%, and a maximum of 2 interventions have been required to achieve closure of the sinus tract orifice during a follow-up ranging from 1 month to 3 years.15,25-30 In the series reported by Kim et al.,28 2 patients required salvage treatment with a cervicotomy. To our knowledge, the cause of failure of endoscopic treatment has never been reported. Sayadi et al.26 and Peireira and Smith29 are the only known groups to have performed a follow-up endoscopy to verify the closure of the sinus tract orifice, and each of these reports describes only 2 patients. Consequently, it is possible that the real rate of recurrence may be higher than actually reported in the other series.15,25,27,28,30 Thus, while our results may appear to be inferior to those reported in the literature, all of our patients underwent follow-up endoscopy, which was not the case in most series reporting endoscopic treatment.15,25,27,28,30 We found (and treated) 3 cases of recurrence during our systematic follow-up endoscopy; in all 3 of these cases, the patients were asymptomatic. Moreover, with a mean follow-up of 23 months, it is improbable that further recurrences will be seen in our series.

We prefer to use the endoscopic treatment approach for this condition whenever possible because it is fast and simple, and it allows us to avoid the risks of open cervical surgery. Complications appear to be very rare, and recurrences can be treated by a second endoscopic cauterization with good long-term outcomes.

Analysis of our results and those reported in the literature has allowed us to identify substantial heterogeneity in the management of fourth branchial pouch sinus tracts. The differences between the 2 main forms of this clinical entity have led us to propose standardized approaches for each. In neonatal cases, the adjacent lesion usually consists of a cyst with its own well-defined wall. These lesions, which are naturally secreting, are seemingly not good candidates for endoscopic management, which is principally used in this situation to exclude and isolate an adjacent lesion associated with the piriform sinus tract. A recurrence of a collection with reopening of the tract seems highly probable. If there is early recurrence following incision of the cyst or noninvolution after antibiotic treatment, then a cervicotomy should be considered. The 3 successful neonatal cases of endoscopic treatment reported herein may have been due to the surrounding infection leading to sclerosis of the cyst wall. Nevertheless, these cases represent the first report to our knowledge of successful endoscopic cauterization in neonates.

On the other hand, in late-onset cases, the poorly defined limits of the lesion and the absence of a cyst wall support an endoscopic approach. In such cases, the cauterization of the superficial tract should not lead to a subsequent relapse unless there is uncontrolled active infection. Based on these observations, we created a therapeutic algorithm, taking into account the age of the patient and the clinical presentation. This study is unique in highlighting these 2 forms of fourth branchial pouch anomalies: (1) a neonatal form that presents with a voluminous cervical mass containing air and possibly requiring a cervicotomy; and (2) a late-onset form in children that presents with abscess formation and for which an endoscopic approach is indicated.

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Correspondence: Nicolas Leboulanger, MD, Service d’Otologie, de Laryngologie et de Rhinologie, et de Chirurgie Cervico-Faciale, Hôpital d’Enfants Armand-Trousseau, 26 Avenue du Dr Netter, 75012 Paris, France (nicolas.leboulanger@trs.aphp.fr).

Author Contributions: Dr Leboulanger had full access to all the data in the study and takes responsibility for the integrity of the data and the accuracy of the data analysis. Study concept and design: Leboulanger, Ruellan, and Roger. Acquisition of data: Leboulanger, Ruellan, Nevoux, Pezzettigotta, Denoyelle, Roger, and Garabedian. Analysis and interpretation of data: Leboulanger, and Ruellan. Drafting of the manuscript: Leboulanger, Ruellan, and Roger. Critical revision of the manuscript for important intellectual content: Nevoux, Pezzettigotta, Denoyelle, Roger, and Garabedian. Statistical analysis: Leboulanger and Ruellan. Administrative, technical, and material support:...
REFERENCES


