Paradoxical Vocal Cord Dysfunction in Juveniles

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Objective: To evaluate demographic and videolaryngoscopic features in a large series of juveniles with paradoxical vocal cord dysfunction (PVCD).

Design: Case series data from videolaryngoscopic tapes retrospectively evaluated in a masked, controlled fashion, and demographic data collected via retrospective medical chart review.

Setting: A tertiary care otolaryngology and speech pathology referral center.

Patients: Twenty-two patients with PVCD aged 18 years and younger diagnosed as having PVCD at The Ohio State University Voice Institute, Columbus.

Main Outcome Measures: Age, sex, social history, and medical history (demographic); epiglottic position, arytenoid and interarytenoid appearance, phase 0 stability, true vocal cord respiratory motion, degree of antero-posterior (AP) constriction, and false vocal cord adduction (videolaryngoscopic).

Results: Of 22 patients, 18 were girls, and 12 had significant social stressors, particularly organized sports. Nineteen patients had posterior laryngeal changes commonly found in gastroesophageal reflux disease. Twelve patients demonstrated abnormal true vocal cord adduction during quiet respiration. Seven patients demonstrated supraglottic anteroposterior constriction and false vocal cord approximation during phonation.

Conclusions: Juvenile PVCD is more common in girls and is associated with social stresses. Anatomic laryngeal changes typically associated with gastroesophageal reflux disease are extremely common in these patients. Juveniles with PVCD frequently demonstrate abnormal true vocal cord adduction during quiet respiration. We recommend that initial evaluation of juvenile patients for possible PVCD be conducted via transnasal fiberoptic laryngoscopy while the patient is asymptomatic, and that strong consideration be given to empiric pharmacological treatment of gastroesophageal reflux disease in juveniles diagnosed as having PVCD.

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Paradoxical Vocal Cord Dysfunction (PVCD) is a recently described syndrome characterized by paroxysmal periods of true vocal cord (TVC) adduction during inspiration, non-phonatory expiration, or both. The term “paradoxical,” with regard to abnormal adduction of TVCs during respiration, was first used in 1978. Concurrently, descriptions of “Munchausen stridor” and “factitious asthma” appeared in the literature in response to the apparent lack of organic pathology associated with, and the possible psychological cause of, this clinical entity. During the past 20 years, the characteristic history, physical examination results, and laboratory findings for PVCD, as well as various treatment options for this disorder, have been the subject of intense investigation. Patient history for PVCD typically consists of sporadic attacks of stridor or wheezing, frequently during activity. This often creates confusion among physicians who care for these patients, and many are diagnosed as having reactive airway disease and undergo extended bronchodilator and corticosteroid therapy. These patients typically have normal pulse oximetry, blood gas values, and chest radiographs, and do not respond to histamine or methacholine challenges. Flow volume loops obtained during attacks typically show a blunted inspiratory component in these patients. Such laboratory values may help differentiate PVCD from asthma, but recent studies documented the presence of asthma in as many as half of all patients with PVCD. Therefore, the diagnostic standard for PVCD is direct observation of TVC adduction during inspiration, expiration, or both during laryngoscopy. By definition, patients must also demonstrate normal TVC abduction.
PATIENTS AND METHODS

From January 1, 1990, to December 31, 1997, 26 juveniles (≥18 years) diagnosed as having PVCD at The Ohio State University Department of Otolaryngology and Voice Institute were referred for evaluation after negative findings on thorough pulmonary workup in The Ohio State University Department of Pulmonary Medicine, the Pulmonary Medicine Division of Columbus Children’s Hospital, or a similar outpatient pulmonary medicine clinic in Ohio. Their initial evaluations at the Voice Institute included a thorough history and complete head and neck examination. During videolaryngoscopy, patients were studied during quiet respiration and multiple task phonation (sustained vowels, musical glides, and ah-ha repeats). We evaluated the presence of normal TVC adduction with maneuvers such as deep nasal inspiration (“sniffing”) to identify the breathing task that best correlated with an open glottis. Images were obtained during sustained vowel production using a stroboscope (model 9100; Kay-Elemetrics, Pine Brook, NJ). All patients were asymptomatic during initial endoscopic evaluation, during which no provocative testing was performed. Endoscopic and videolaryngostroboscopic recordings were taken after patients had adapted to the equipment or had received topical anesthetic treatment with lidocaine hydrochloride (10%) spray to minimize the involuntary gag reflex.

For the purpose of this study, a retrospective review of our series of 26 juveniles with PVCD was performed. Initial endoscopy videotapes for these patients and 7 normal examination tapes that served as internal controls were randomized, copied onto a master videotape, and blindly evaluated by 3 experienced clinicians (K.T., M.D.T., and L.A.F.). Evaluators were informed that the master tape contained randomized endoscopic videotapes from patients with PVCD and healthy volunteers. Data collected during tape review included epiglottic position, arytenoid anatomic description, interarytenoid space evaluation, TVC motion during respiration, degree of anteroposterior (AP) constriction of the supraglottic larynx during examination (estimated and reported as a percentage of total supraglottic AP dimension), degree of false vocal cord (FVC) adduction during phonation (recorded on a 3-point scale: 1 indicates no or minimal adduction; 2, moderate adduction; and 3, complete false vocal fold approximation), and phase 0 stability (either stable or unstable).

If more than 1 evaluator thought that a patient’s taped examination was inadequate for data collection, that patient was excluded from further evaluation and was not entered into our final data summary table. Endoscopic videotapes were considered inadequate for review in 4 cases; therefore, our review includes data from only the remaining 22 patients.

For subjective data to be included in the analysis, at least 2 of 3 evaluators were required to report the same finding for each variable. Moreover, if 2 or more evaluators listed a variable as “normal” or reported mixed (normal and abnormal) observations for a particular variable, the patient would receive a score of normal for that variable. If 2 or more evaluators reported a numerical score greater than 0% for supraglottic AP constriction or greater than 1 for degree of FVC adduction, these scores were averaged to obtain a single score for inclusion in the data summary table. Finally, social and medical histories were collected from a retrospective medical chart review and included in the final data presentation. The Fisher exact test was used for statistical analysis of the degree of relationship between any 2 variables. $P ≤ .05$ was accepted as the minimal level of statistical significance.

Numerous investigators have reported attempts to provoke this classic finding in the office by stimulating a typical attack using exercise tasks or exposure to chemical irritants. The cornerstones of therapy for PVCD are laryngeal control therapy via speech and breathing techniques and psychological counseling. However, many other treatments have been reported, including heliox, hypnosis, botulinum toxin, sedation, coughing, panting, and even intubation and tracheotomy. The efficacy of these treatments, however, has not been systematically evaluated.

As articles describing various clinical features of PVCD have accumulated in the literature, a view has emerged that PVCD may represent a spectrum of disease with psychological and physiological causes. The psychological characteristics of the typical patient have been well documented and are discussed elsewhere. The typical patient heretofore described is a woman, aged 20 to 40 years, overweight and with significant stress problems, and who has some knowledge of medical care. Physiological abnormalities associated with PVCD include asthma, cystic fibrosis, and gastroesophageal reflux disease (GERD). Recently, classification systems have been proposed in an attempt to define the subgroups of PVCD.

Most clinical data for PVCD have been reported in the context of case reports and occasionally case series. However, there has been little effort to study juvenile patients separately from adults. A literature review revealed only 3 articles that specifically report series of juveniles with PVCD, with a total of 15 patients described. Juveniles with PVCD might be different from adults with PVCD with regard to coincident pathology and, therefore, might compose a different subgroup of a complex disease spectrum.

In light of the relative paucity of literature concerning juveniles with PVCD, further clinical data are necessary to characterize this disorder and its associations with factors such as GERD and social stressors in this patient population. At The Ohio State University Voice Institute, Columbus, we collected data from a large series of juveniles with PVCD. The primary goals of this study were to investigate (1) the utility of videolaryngoscopy in asymptomatic juvenile patients with known PVCD, (2) the presence or absence of laryngeal abnormalities associated with PVCD in our series of juveniles with this disorder, and (3) demographic features of juveniles with PVCD.

Of 26 tapes from juvenile patients with documented PVCD, 22 were considered adequate by the 3 evaluators. The 4 inadequate tapes were all recorded via oral endoscopy.
Demographic findings for the remaining 22 patients are summarized in Table 1. The average age of juvenile patients with PVCD was 14.5 years (range, 9.0-18.0 years); 18 (82%) were girls and 4 (18%) were boys. Twelve patients with PVCD (55%) had a history of important social stresses: 9 were high school athletes who played competitive organized sports and 3 were heavily involved in other extracurricular activities.

Two patients were diagnosed early in their disease course as having asthma. These and 2 other patients were treated with trials of bronchodilators. None of the remaining 18 patients had a significant medical history, and all were nonsmokers and nondrinkers.

Videolaryngoscopy findings for the 22 patients with PVCD are summarized in Table 2. The method of laryngoscopy seemed to affect laryngeal examination findings. Eight (36%) of 22 patients had abnormal epiglottic positioning during evaluation: all were described as having a posteriorly displaced epiglottis, and 7 (88%) were evaluated via oral endoscopy. There was a statistically significant concordance between oral laryngoscopy and observation of a posteriorly displaced epiglottis (P = .001).

With regard to posterior glottic changes, 17 patients (77%) with PVCD demonstrated arytenoid edema, and 19 (86%) had interarytenoid abnormalities (Figure 1). Of 19 patients with interarytenoid findings, evaluators described the interarytenoid space as edematous in 11 patients (50%) and as demonstrating frank pachyderma in 8 (36%).

Abnormal laryngeal physiological findings were observed in most videolaryngoscopy examinations of symptomatic juveniles with PVCD. Five patients (23%) with PVCD demonstrated an unstable phase 0 on stroboscopy. Using the Fisher exact test, there was no correlation between an unstable phase 0 and either posterior glottic changes or abnormal respiratory TVC motion. Twelve patients (55%) demonstrated abnormal TVC adduction during quiet, asymptomatic respiration (see Figure 2 for an example image from a test videotape). Nine patients (41%) had a significant degree of AP constriction of the supraglottis; among this subgroup, the average degree of AP constriction was 39.7%. Ten patients (45%) demonstrated abnormal FVC adduction during examination, 7 (70%) of whom were also judged to have an estimable degree of AP constriction (average, 37.4%). The relation between AP constriction and FVC adduction was statistically significant (P = .03) (see Figure 3 for an example image from a videotape).

Our series of juvenile patients with PVCD mostly comprised teenage girls (82%). Our finding of a female sex

<table>
<thead>
<tr>
<th>Patient No.</th>
<th>Sex/Age, y</th>
<th>Social Factors</th>
<th>Illnesses</th>
<th>Allergies</th>
<th>Medications Used</th>
<th>Chief Complaint</th>
</tr>
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<tbody>
<tr>
<td>1</td>
<td>F/16</td>
<td>Volleyball, basketball</td>
<td>None</td>
<td>None</td>
<td>None</td>
<td>SOB during sports</td>
</tr>
<tr>
<td>2</td>
<td>F/15</td>
<td>Volleyball, basketball</td>
<td>None</td>
<td>None</td>
<td>None</td>
<td>SOB during basketball</td>
</tr>
<tr>
<td>3</td>
<td>F/14</td>
<td>No activities</td>
<td>None</td>
<td>None</td>
<td>None</td>
<td>Intermittent SOB</td>
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<td>F/16</td>
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<td>None</td>
<td>None</td>
<td>Intermittent SOB, “wheezing”</td>
</tr>
<tr>
<td>5</td>
<td>F/13</td>
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<td>None</td>
<td>None</td>
<td>None</td>
<td>Intermittent SOB</td>
</tr>
<tr>
<td>6</td>
<td>F/14</td>
<td>Soccer, track</td>
<td>Environmental allergies</td>
<td>None</td>
<td>None</td>
<td>SOB, wheezing with all sports</td>
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<tr>
<td>7</td>
<td>M/17</td>
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<td>None</td>
<td>None</td>
<td>Albuterol (trial) prn</td>
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</tr>
<tr>
<td>8</td>
<td>F/18</td>
<td>Track, tennis</td>
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<td>None</td>
<td>Intermittent wheezing</td>
</tr>
<tr>
<td>9</td>
<td>F/15</td>
<td>Basketball, track</td>
<td>Acne</td>
<td>None</td>
<td>Minocycline</td>
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</tr>
<tr>
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<td>SOB with sports practice</td>
</tr>
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<td>None</td>
<td>None</td>
<td>SOB with “stress”</td>
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<tr>
<td>12</td>
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<td>None</td>
<td>None</td>
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<tr>
<td>13</td>
<td>F/14</td>
<td>“Extracurricular activities”</td>
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<td>None</td>
<td>None</td>
<td>Not recorded</td>
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<td>14</td>
<td>F/15</td>
<td>Lifeguard, choir</td>
<td>Asthma, vasomotor rhinitis</td>
<td>Narcotics, neosporin</td>
<td>Triamcinolone acetonide, nedocromil sodium</td>
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<td>15</td>
<td>F/15</td>
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<td>None</td>
<td>None</td>
<td>Intermittent SOB</td>
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<td>None</td>
<td>SOB with lacrosse</td>
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<td>17</td>
<td>F/17</td>
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<td>None</td>
<td>None</td>
<td>Intermittent SOB</td>
</tr>
<tr>
<td>18</td>
<td>M/9</td>
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<td>None</td>
<td>None</td>
<td>None</td>
<td>Intermittent SOB</td>
</tr>
<tr>
<td>19</td>
<td>F/13</td>
<td>“Exercise enthusiast”</td>
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<td>None</td>
<td>Albuterol sulfate (trial) prn</td>
<td>SOB, stridor with exercise</td>
</tr>
<tr>
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<td>F/14</td>
<td>No activities</td>
<td>Environmental allergies, asthma</td>
<td>None</td>
<td>Several inhalers (trial) prn</td>
<td>Intermittent stridor, occasionally with SOB</td>
</tr>
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<td>None</td>
<td>Allergy shots, prednisone (trial), antihistamines prn</td>
<td>Intermittent SOB</td>
</tr>
<tr>
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<td>M/11</td>
<td>No activities</td>
<td>None</td>
<td>None</td>
<td>None</td>
<td>Intermittent SOB</td>
</tr>
</tbody>
</table>

*PVCD indicates paradoxical vocal cord dysfunction; SOB, shortness of breath; and prn, as occasion requires.
predominance and the high incidence (55%) of significant social stresses are consistent with previous demographic findings\(^9,11\) for adult patients with PVCD.

Analysis of laryngoscopic data revealed that the endoscopic technique had a statistically significant effect on the appearance of supraglottic structures. Seven of 8 patients judged to have a posteriorly displaced epiglottis were evaluated via oral endoscopy. Only 1 oral endoscopy was scored as normal with regard to static appearance of the supraglottis. However, dynamic supraglottic abnormalities, as measured by abnormal AP constriction and FVC adduction, did not relate significantly to oral endoscopy. These results indicate that oral endoscopy may offer a different perspective on the static appearance of the supraglottis, which may interfere with interpretation of endoscopic data. These variables have not been discussed previously, and we recommend that clinicians use oral endoscopy only if the patient is intolerant of nasal endoscopy so as to avoid this possibly confounding factor.

A high proportion of our juvenile patients with PVCD demonstrated posterior glottic changes typically seen in GERD. Twenty-one (95%) of 22 patients, all nonsmokers, demonstrated notable arytenoid edema and interarytenoid edema or pachyderma on endoscopic examination of the posterior glottis; 36% demonstrated frank interarytenoid pachyderma, which is typically associated with chronic reflux laryngitis.\(^19\) These abnormalities would not be expected in healthy adolescent larynges, and they support the association between PVCD and GERD reported by multiple investigators.\(^1\)\(^6\),\(^\)\(^7\) Paradoxi-
Paradoxical vocal cord dysfunction may represent a low-grade form of laryngospasm in response to irritation from reflux events, which have been documented to cause vocal cord adduction and apnea in infants. However, definitive testing using techniques such as esophageal pH probe evaluation would be necessary to determine the true incidence of GERD in juveniles with PVCD. Investigators previously proposed that patients with PVCD be subdivided into cases secondary to physiological causes, such as neuropathologic disorders or GERD, and those secondary to psychological causes (ie, malingerers and conversion disorder patients). The prevalence of posterior laryngeal changes—classically seen in GERD—in the present sample supports the notion that PVCD can occur secondary to disease processes and not solely secondary to psychological factors, as had been proposed in early studies.

Another frequent (55%) observation was of abnormal TVC adduction during quiet respiration in our initial videotapes of asymptomatic juveniles evaluated for PVCD. This observation is important because many articles report that provocative testing is required for definitive diagnosis of PVCD. Newman and colleagues reported a large series of 95 patients with PVCD and found a similar proportion of TVC adduction in asymptomatic patients, but the authors did not examine or report juvenile patients separately. The few other articles specifically reporting juveniles with PVCD do not comment on this finding. The results of the current study suggest that PVCD may be diagnosed by laryngoscopic evaluation of asymptomatic patients, reserving exercise challenge testing for those with equivocal findings. However, the high association between asthma and PVCD reported by Newman et al and other investigators prompts a consideration of methacholine challenge testing in order to rule out concomitant reactive airway disease in patients with PVCD.

Another observation made by evaluators is the presence of FVC adduction and AP constriction during laryngoscopic evaluation of patients with PVCD. Although less than half of juvenile patients demonstrated these abnormalities (45% and 41%, respectively), the correlation between these 2 findings was high, since 70% of those with notable FVC adduction also demonstrated significant AP constriction. These findings may indicate a subset of juvenile patients with PVCD who have abnormal laryngeal physiological findings. Although supraglottic constriction may be achieved voluntarily, the muscles involved are also mediated by vagal centers that control involuntary vocal cord movement. Vagal irritability may also account for the 23% incidence of an unstable phase 0 on stroboscopy in this sample. This observation did not correlate significantly with other laryngoscopic examination findings. Together, these observations suggest that a subset of patients with PVCD has poor laryngeal muscle control or lack of coordination and warrants consideration of a neuropathologic cause for PVCD, despite the absence of neurologic disease in this case series.

Because supraglottic muscles are under partial voluntary control, it is equally possible that these patients represent a subgroup with PVCD associated with psychological factors. Previous studies have proposed a link between psychological illness or stressors and PVCD. Maschka et al further subdivide this group into malingerers patients motivated by secondary gain, and conversion disorder patients who are overwhelmed by social stresses. In our series, the majority of juvenile patients diagnosed as having PVCD (55%) identified important social stresses, usually high school sports.

For these 22 juveniles with PVCD, demographic findings were similar to those previously reported in the literature for adults with PVCD. A high incidence of abnormal laryngeal physiological findings—including changes typical of GERD, an unstable phase 0, TVC respiratory adduction while asymptomatic, and supraglottic constriction—was observed on videolaryngoscopy. These findings support the notion that PVCD represents a complex, heterogeneous disorder with multiple physiological abnormalities that are difficult to attribute to its initially reported psychological cause.

CONCLUSIONS

In this case series of juveniles with PVCD, 18 of 22 were girls, and the majority reported significant social stresses such as competitive sports activities. These findings support a previously reported association between important social stresses and diagnosis of PVCD in adults. In this investigation, oral laryngoscopy distorted the appearance of epiglottic position; therefore, nasal laryngoscopy should be regarded as superior and should be the technique-of-choice for evaluating these patients. The majority of juveniles with PVCD demonstrated abnormal TVC adduction during quiet respiration while asymptomatic, without provocative testing. The diagnosis, therefore, may be established for most juvenile patients with PVCD in an office setting via transnasal flexible fiber-optic laryngoscopy, without resorting to provocative testing or sophisticated stroboscopic evaluation. However, there is a role for subsequent methacholine challenge testing, even after diagnosis of PVCD, because many investigators have demonstrated that asthma may be present in patients with PVCD.
There was a strong association between PVCD and posterior laryngeal anatomic abnormalities in this patient population, including arytenoid edema and interarytenoid pachyderma, which are laryngeal findings typical of GERD. Although definitive testing would be necessary to prove the presence of GERD, an empirical trial of antireflux pharmacological therapy is warranted on the basis of these commonly identified posterior laryngeal changes in juveniles with PVCD, in conjunction with the usual therapeutic interventions of speech therapy and psychological counseling. A subset of these patients demonstrates abnormal laryngeal physiological findings with poor laryngeal control, which supports the concept that PVCD is a complex disorder that has psychological and physiological components.

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