Objective: To evaluate the prevalence of sleep-disordered breathing (SDB) and/or obstructive sleep apnea (OSA) in the population with nonsyndromic cleft palate.

Design: Retrospective medical record review of symptoms of SDB and/or OSA and results of polysomnography (PSG) studies.

Setting: The craniofacial clinic of a tertiary pediatric hospital.

Patients: A total of 459 patients, with an additional 48 patients with Pierre Robin syndrome, met inclusion criteria.

Main Outcome Measures: Medical records from January 1, 2005, through July 31, 2009, were reviewed for demographic data, SDB symptoms, surgical procedures, and PSG results.

Results: Of the 459 patients, 172 (37.5%) had symptoms of SDB and 39 (8.5%) had PSG-diagnosed OSA. Forty-six patients underwent 1 or more PSGs, with results of 49 of the 59 studies (83.1%) being positive for OSA. Surgical procedures to address SDB and/or OSA were undertaken in 89 patients (51.7%), with combined tonsillectomy and adenoidectomy the most common procedure (44.9%). An additional 48 patients who met the inclusion criteria with a diagnosis of Pierre Robin syndrome were also identified. In this population, 35 patients (72.9%) had symptoms of SDB and/or OSA.

Conclusions: An increased prevalence of SDB and/or OSA exists in the population with cleft palate, with an even greater prevalence in patients with Pierre Robin syndrome. Definitive diagnosis of OSA by PSG is underused. We suggest that surgical management of SDB and/or OSA be followed by PSG to demonstrate resolution or persistence of symptoms to ensure appropriate further management.

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SLEEP-DISORDERED BREATHING (SDB) is a spectrum of diseases ranging from primary snoring to obstructive sleep apnea (OSA). Primary snoring, the least severe of the spectrum, consists of nightly or frequent snoring without apnea, hypoventilation, or sleep fragmentation and does not have any associated daytime symptoms. Upper airway resistance syndrome constitutes the middle of the spectrum and is accompanied by increased respiratory effort and daytime symptoms but is not associated with gas-exchange abnormalities. Obstructive sleep apnea constitutes the most severe end of the spectrum (see the article by Reveni et al for a more comprehensive review of the spectrum of SDB). In the pediatric population, OSA is defined as a “disorder of breathing during sleep characterized by prolonged partial upper airway obstruction and/or intermittent complete obstruction that disrupts normal ventilation during sleep and normal sleep patterns.” Symptoms include habitual snoring, labored breathing during sleep, observed apnea, restless sleep, dia- phoresis, enuresis, cyanosis, excessive daytime sleepiness, and behavior or learning problems (see the Clinical Practice Guideline of the American Academy of Pediatrics for a comprehensive review of OSA in pediatrics). Risk factors for OSA include adenotonsillar hypertrophy, obesity, craniofacial anomalies, and neuromuscular disorders. Accurate diagnosis is required not only to ensure proper treatment but also to prevent possible complications, such as systemic hypertension, pulmonary hypertension, failure to thrive, neurocognitive impairment, and behavioral problems. With regard to diagnosis of OSA, multiple studies have shown that OSA questionnaires and history and physical examination only are not reliable. The criterion standard for diagnosis is polysomnography (PSG), which can be performed on children of any age. The prevalence of OSA in the general pediatric population is approximately 2% to 3%, with 3% to 12% having primary snoring. As mentioned, the presence of craniofacial anomalies increases the risk of OSA, and those individuals with cleft palate with or without a cleft lip (CPL) fall into this high-risk population.
Cleft palate with or without a cleft lip is a relatively common birth defect, with an incidence of 0.69 to 2.51 per 1000 births worldwide and 1 in 680 live births in the United States each year. It results from failure of the medial nasal process to contact or to maintain contact with the lateral nasal and maxillary processes between the sixth and ninth weeks of prenatal development. Epidemiologic studies indicate that 70% of clefts occur as isolated malformations and 30% occur in the context of a syndrome, as a chromosomal abnormality, or in association with multiple congenital anomalies of unknown origin. In children with CP/L, the associated facial features include midface hypoplasia and a retrognathic maxilla, resulting in reduced upper airway dimensions that persist even after surgical correction. These anatomical differences may place these patients at increased risk of SDB, with studies estimating the risk of OSA to be between 22% and 65% in infants and children with CP/L.

In addition to being at risk of OSA because of congenital anatomical differences, patients with CP/L are at additional increased risk secondary to surgical procedures performed to help correct or repair the cleft and to improve speech. Approximately 10% to 53% of children with CP/L require surgery for velopharyngeal insufficiency (VPI), usually through procedures such as palatoplasty, pharyngeal flap, and sphincter pharyngoplasty. These procedures often decrease the cross-sectional area of the airway, and multiple studies have concluded that these procedures increase the risk of OSA.

The goals of this study are to help determine the prevalence of SDB and/or OSA in the population with nonsyndromic cleft palate through a retrospective review of patients from the Children’s Hospital of Pittsburgh of University of Pittsburgh Medical Center Craniofacial Clinic from January 1, 2005, through July 31, 2009. Inclusion criteria for the study included nonsyndromic patients with CP/L who had undergone cleft repair. Patients with a diagnosis of Pierre Robin syndrome (PRS) were analyzed separately. Patients and/or families were routinely questioned directly about symptoms of snoring and apneic episodes, with most patients and families also filling out a review-of-symptoms questionnaire at every visit that included these symptoms. Clinical notes and questionnaires were used to ascertain the presence of SDB symptoms. Patients who had a medically confirmed diagnosis of a congenital syndrome were excluded from the study. A total of 36 different congenital syndromes were identified in patients from the database.

Medical records were reviewed for the following data: type of cleft palate (cleft palate and lip vs cleft palate only and bilateral vs unilateral); age at time of diagnosis of SDB; presence of SDB symptoms before, after, or both with regard to timing of cleft repair; surgical procedures to address SDB; age when surgical procedure(s) performed; and PSG (number of studies and results). The average and median ages were determined for the diagnosis of SDB, performance of surgical procedures to address SDB, and performance of PSG.

The PSG results were reviewed when available and used as the criterion standard for the diagnosis of OSA. The data point obtained from the PSG used to diagnosis OSA was the apnea-hypopnea index (AHI). The AHI values were graded into severity levels according to variables described by Muniz et al. The AHI values less than 1 were considered normal; 1 to 4.9, mild OSA; 5.0 to 14.9, moderate OSA; and greater than 15, severe OSA.

RESULTS

A total of 459 patients met the inclusion criteria, and an additional 48 patients met inclusion criteria but also had a diagnosis of PRS. Of those patients without PRS, 172 (37.5%) had symptoms of SDB that included snoring and/or apneic pauses in breathing during sleep. The average age at time of diagnosis of SDB was 5.2 years (median, 4 years; range, 0-18 years). Forty-six of the 172 SDB patients underwent PSG, with 39 (22.7% of SDB patients and 8.5% of all patients) having at least 1 study that met the criteria for diagnosis of OSA. The severity level of OSA was determined by the AHI and was analyzed and broken down by patient age at the time of the initial PSG (Table 1).

Further analysis of patients with cleft palate, including type of cleft (cleft palate and cleft lip vs cleft palate

### METHODS

Institutional review board approval was obtained to search the Craniofacial Clinic database that contained information for 757 patients seen in the clinic from January 1, 2005, through July 31, 2009. Inclusion criteria for the study included nonsyndromic patients with CP/L who had undergone cleft repair. Patients with a diagnosis of Pierre Robin syndrome (PRS) were analyzed separately. Patients and/or families were routinely questioned directly about symptoms of snoring and apneic episodes, with most patients and families also filling out a review-of-symptoms questionnaire at every visit that included these symptoms. Clinical notes and questionnaires were used to ascertain the presence of SDB symptoms. Patients who had a medically confirmed diagnosis of a congenital syndrome were excluded from the study. A total of 36 different congenital syndromes were identified in patients from the database.

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only and bilateral vs unilateral cleft), showed similar results to the population as a whole with regard to the percentage with SDB symptoms, number of patients undergoing surgery to address SDB, and PSG results (Table 2).

Medical records were reviewed to determine the timing of the diagnosis of SDB with regard to surgical repair of the cleft palate. Eight patients (4.7%) had symptoms present only before surgical repair, 118 patients (68.6%) after surgical repair, and 29 patients (16.9%) with symptoms present before and after repair. A separate category of 17 patients (9.9%) was created for those with appearance of SDB after cleft repair and, more specifically, those who developed symptoms after undergoing a pharyngeal flap procedure to treat VPI. Of the 459 patients, 117 (25.5%) underwent a pharyngeal flap procedure for VPI, and 79 (67.5%) of them had had SDB at some point. Table 4 presents the timing of presentation of SDB by age group.

Surgical procedures directed at correcting or improving SDB were undertaken for 89 patients (51.7% of patients with SDB). The most common procedure for all age groups except that of patients older than 10 years was combined tonsillectomy and adenoidectomy (which also included tonsillectomy with partial adenoidectomy), with 44.9% of those receiving surgery undergoing this procedure. The other common procedures included pharyngeal flap takedown (21.3%), adenoidectomy or partial adenoidectomy only (13.5%), tonsillectomy only (9.0%), and other procedures, such as tracheostomy (2 procedures), septoplasty or rhinoplasty (5 procedures), and maxillary distraction (3 procedures). A total of 11 patients underwent 2 different procedures. Age distribution and surgical procedures are listed in Table 5.

There was also a population of patients with PRS who met all the other inclusion criteria. Of the 48 patients in this subpopulation, 35 (72.9%) had SDB. Four of these patients underwent a total of 18 PSGs, with results of 6 studies being abnormal (33.3%). An almost equal numbers of these patients had SDB symptoms before and after surgical correction of the cleft (14 [40.0%]) or only

<table>
<thead>
<tr>
<th>Cleft Subgroup</th>
<th>Total No. of Patients</th>
<th>SDB (37.5%)</th>
<th>Surgery (51.7%)</th>
<th>PSG (26.7%)</th>
<th>Positive PSG Results (84.8%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>All</td>
<td>459</td>
<td>172</td>
<td>89</td>
<td>46</td>
<td>39</td>
</tr>
<tr>
<td>Bilateral</td>
<td>82 (17.9)</td>
<td>35 (42.7)</td>
<td>17 (48.6)</td>
<td>7 (20.0)</td>
<td>5 (71.4)</td>
</tr>
<tr>
<td>Unilateral</td>
<td>377 (82.1)</td>
<td>137 (36.3)</td>
<td>72 (25.6)</td>
<td>39 (28.5)</td>
<td>34 (87.2)</td>
</tr>
<tr>
<td>Lip and palate</td>
<td>303 (66.0)</td>
<td>104 (34.3)</td>
<td>56 (53.8)</td>
<td>27 (26.0)</td>
<td>22 (81.5)</td>
</tr>
<tr>
<td>Palate only</td>
<td>156 (34.0)</td>
<td>68 (43.6)</td>
<td>33 (48.5)</td>
<td>19 (27.9)</td>
<td>17 (89.5)</td>
</tr>
</tbody>
</table>

Abbreviations: PSG, polysomnography; SDB, sleep-disordered breathing.

<table>
<thead>
<tr>
<th>Cleft Subgroup</th>
<th>Total No. of Patients With SDB</th>
<th>0-2 (32.0%)</th>
<th>3-5 (26.2%)</th>
<th>6-10 (28.5%)</th>
<th>&gt;10 (23.4%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>All</td>
<td>172</td>
<td>55 (32.0%)</td>
<td>45 (26.2%)</td>
<td>49 (28.5%)</td>
<td>23 (13.4%)</td>
</tr>
<tr>
<td>Bilateral</td>
<td>35</td>
<td>8 (32.0%)</td>
<td>9 (36.0%)</td>
<td>9 (25.7%)</td>
<td>8 (22.9%)</td>
</tr>
<tr>
<td>Unilateral</td>
<td>137</td>
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<td>36 (26.3%)</td>
<td>39 (28.5%)</td>
<td>15 (10.9%)</td>
</tr>
<tr>
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<td>104</td>
<td>30 (28.6%)</td>
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<td>32 (30.8%)</td>
<td>17 (16.3%)</td>
</tr>
<tr>
<td>Palate only</td>
<td>68</td>
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<td>17 (25.0%)</td>
<td>6 (8.8%)</td>
</tr>
</tbody>
</table>

Abbreviation: SDB, sleep-disordered breathing.

<table>
<thead>
<tr>
<th>Age, y</th>
<th>Total No. of Patients With SDB</th>
<th>Before Repair</th>
<th>After Repair</th>
<th>Before and After Repair</th>
<th>After Pharyngeal Flap Procedure</th>
</tr>
</thead>
<tbody>
<tr>
<td>0-2</td>
<td>55</td>
<td>6 (10.9)</td>
<td>27 (49.1)</td>
<td>22 (40.0)</td>
<td>0</td>
</tr>
<tr>
<td>3-5</td>
<td>45</td>
<td>1 (2.2)</td>
<td>31 (68.9)</td>
<td>6 (13.3)</td>
<td>7 (15.6)</td>
</tr>
<tr>
<td>6-10</td>
<td>49</td>
<td>1 (2.0)</td>
<td>37 (75.5)</td>
<td>1 (2.0)</td>
<td>10 (20.4)</td>
</tr>
<tr>
<td>&gt;10</td>
<td>23</td>
<td>0</td>
<td>23 (100)</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>Total</td>
<td>172</td>
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</table>

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after (13 [37.1%]). Six patients (17.1%) had symptoms before cleft repair only. Surgical procedures to address SDB in this subpopulation included tracheostomy (30.8%; average age, 0 years; median age, 0 years), mandibular distraction (15.4%; average and median ages, 9 months), adenoidectomy (7.7%; average and median ages, 5 years); pharyngeal flap takedown (15.4%; average age, 19 years; median age, 19 years), tonsillectomy (7.7%; average age, 8 years; median age, 8 years), maxillary distraction (7.7%; average age, 10 years; median, 10 years), tongue-lip adhesion (7.7%; average age, 0 years; median age, 0 years), and tonsillectomy with adenoidectomy (7.7%; average age, 7 years; median age, 7 years). Two patients each underwent 2 different procedures, with the remaining patient undergoing only 1 procedure. Tables 6, 7, and 8 review the onset and presence of symptoms, PSG results, and surgical procedures by age for patients with PRS.

The overall prevalence of SDB in the population with cleft palate was 37.5%, significantly higher than the 3% to 12% in the healthy pediatric population.5,6 The prevalence of PSG-confirmed OSA was 8.5%, approximately 3 times the healthy pediatric population prevalence of 2% to 3%.3,4 The SDB prevalence in this study is consistent with the range of 22% to 65% reported in other studies.7,14,15 The youngest age group, 0 to 2 years old, had the largest number of patients diagnosed as having SDB; however, this group had the fewest patients undergoing PSG. All the patients who underwent PSG from this age group had abnormal results. One patient with an abnormal initial PSG result then underwent adenoidectomy and had a subsequent normal PSG result (hence the 1 normal PSG result listed in Table 1). As the age increased, there was a

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trend for a decreasing number of positive PSG results and a corresponding decrease in the percentage of patients with at least 1 positive PSG result. The rationale for determining which patients underwent PSG vs those who did not was not clearly identifiable from the retrospective nature of the study.

For all age groups, onset of symptoms occurred predominantly after surgical repair of the cleft palate defect. The youngest age group had almost equal distribution between onset after surgical repair and presence of symptoms before and after repair. For the other age groups, there was a marked preponderance for postrepair onset of symptoms. In the postrepair population, a proportion of patients had onset of symptoms after undergoing surgical treatment of VPI with a pharyngeal flap procedure. Of the 459 patients, a total of 117 patients underwent at least 5 PSGs when they underwent decannulation after tracheostomy.

Our reported prevalence of SDB by age group may not be an entirely accurate representation of the actual onset or presence of symptoms because of the nature of the study. This retrospective medical record review is limited because of the nature of the medical records themselves. The electronic medical records used in this study did not extend back farther than 2002. Older patients may have had symptoms before those reported in this study, but documentation was unavailable for those earlier periods. In addition, there are some patients who transferred care from other locations, therefore masking the true age at onset of symptoms if they occurred before being seen in our clinic. Having considered these limitations, the results still point to a much higher incidence of SDB after surgical correction. This is particularly evident in the youngest population, for whom we have the most complete medical records.

In the pediatric population, combined tonsillectomy and adenoidectomy are the most common surgical procedures used to address SDB. Tonsillectomy and adenoidectomy were also the most common procedures performed on the patients with clefts, although a partial instead of a full adenoidectomy was frequently performed to prevent future VPI. The next most common procedure was pharyngeal flap takedown, which as expected was performed mostly in older patients. Other procedures such as adenoidectomy (or partial adenoidectomy), tracheostomy, and septoplasty or rhinoplasty were also fairly common. All the tracheostomies were performed on newborns, and maxillary osteotomies or distractions were performed on the oldest patients, with an average age at time of maxillary distraction of 12.5 years (median, 12 years). The success of the individual surgical procedures cannot be objectively measured because of the nature of the medical records used in this study.

Patients with PRS have been reported to have an increased incidence of SDB and/or OSA. In this study, of the 48 patients with PRS, 35 (72.9%) had symptoms of SDB. Interestingly, only a small percentage underwent PSG (5 [14.3%] of SDB patients). Two of these patients underwent at least 5 PSGs when they underwent decannulation after tracheostomy.

Patients with PRS also underwent a different set of surgical procedures for SDB. Four patients (30.8% of all PRS patients with SDB) underwent tracheostomies as newborns. The next most common operation was mandibular distraction (3 patients, 15.4% of surgical patients), with only 1 to 2 patients each having tonsillectomy, adenoidectomy, tonsillectomy and adenoidectomy, pharyngeal flap takedown, or maxillary distraction. This was distinctly different from the surgical procedures used in the CPL population as a whole and reflects the specific anatomical deformities (eg, micrognathia and possible severe airway obstruction) present in the PRS subpopulation.

In conclusion, SDB and OSA are more common in the population with cleft palate than in the general pediatric population, and the true prevalence is probably underreported. The prevalence is increased to a greater extent in those patients with PRS. Direct questioning regarding signs and symptoms of SDB and/or OSA should be an integral part of every office visit for these patients. Polysomnography should be considered for any patient with

Table 8. Surgical Procedures to Address SDB in Patients With PRS According to Age at Time of Surgerya

<table>
<thead>
<tr>
<th>Age, y</th>
<th>Total No. of Patients With SDB</th>
<th>Patients Undergoing Surgery</th>
<th>Tracheostomy</th>
<th>Mandibular Distraction</th>
<th>Adenoidectomy</th>
<th>Pharyngeal Flap Procedure</th>
<th>Tonsillectomy</th>
<th>Tonsillectomy and Adenoidectomy</th>
<th>Maxillary Distraction</th>
<th>Tongue-Lip Adhesion</th>
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<tr>
<td>0-2</td>
<td>23</td>
<td>7 (30.4)</td>
<td>4 (57.1)</td>
<td>2 (28.6)</td>
<td>0</td>
<td>0</td>
<td>0</td>
<td>0</td>
<td>0</td>
<td>1 (14.3)</td>
</tr>
<tr>
<td>3-5</td>
<td>6</td>
<td>1 (16.7)</td>
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<td>0</td>
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<tr>
<td>6-10</td>
<td>4</td>
<td>3 (75.0)</td>
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<td>0</td>
<td>0</td>
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<td>&gt;10</td>
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<td>2 (100)</td>
<td>0</td>
<td>0</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>Total</td>
<td>35</td>
<td>13 (37.1)</td>
<td>4 (30.8)</td>
<td>2 (15.4)</td>
<td>1 (7.7)</td>
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</tr>
</tbody>
</table>

Abbreviations: PRS, Pierre Robin syndrome; SDB, sleep-disordered breathing.

a Two patients underwent 2 procedures each.
positive symptoms and should also be conducted after any surgical intervention to document improvement or resolution of symptoms. Further studies are needed to better understand the true extent of SDB and/or OSA in this patient population as well as the most appropriate surgical and/or medical treatments.

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REFERENCES


