EXTERNAL compression of the trachea from cardiovascular anomalies often causes mechanical upper airway obstruction in children. The complete vascular ring accounts for 85% of vascular tracheobronchial compressions. Tracheal compression by the innominate artery is the most common of the compression syndromes caused by incomplete vascular rings. We report six such patients managed at our hospital over the past five years.

Gross and Neuhauser were the first to associate respiratory difficulties with innominate artery compression of the trachea. Although Gross later reported several additional cases of tracheal compression by the innominate artery, the prevalence of this entity was not widely appreciated. Fearon and Shortreed in 1963 were the first endoscopically to document congenital cardiovascular anomalies that cause tracheal compression. In addition, they coined the term "reflex apnea," which they defined as a reflex respiratory arrest initiated by irritation of the area of tracheal compression either by a bolus of food or mechanical stimulation. Although they were unable to explain the mechanism of this reflex, Fagenholz and his coworkers were able to define a phenomenon of increased respiratory drive and laryngeal chemoreflex with apnea independent of peripheral chemoreceptor activity.

EMBRYOLOGY AND ANATOMY

The innominate artery is formed from the embryonic ventral root between the fourth and sixth branchial arches. At the end of the seventh and eighth weeks of fetal life, the heart descends into the chest and the origin of the innominate artery ascends towards the origin of the left
common carotid artery. Generally, the artery comes to lie directly over the trachea, approximately 1 cm. above the carina (Figure 1).

**PATHOPHYSIOLOGY**

An anomalous origin of the innominate artery was first suggested by Gross in 1948, yet Mustard and his coworkers demonstrated that this is true in only 10% or fewer of cases. In fact, the lesion consists of an innominate artery with the vessel arising from the aortic arch and, in turn, creating a sling which compresses the anterior tracheal wall. Tracheal compression then results in ciliary immotility and subsequent inability adequately to clear the airway of secretions. This then tends to increase and thicken the secretions, resulting in obstruction of airflow and inflammation with clinical presentation of a croup-like syndrome.

**PRESENTING SYMPTOMS**

Our six patients included three boys and three girls. The average age at the time of diagnosis was eight months, with the youngest being diagnosed at three months and the oldest at 22 months (Table I). Five of the six patients (80%) had a history of at least one episode of respiratory distress with cyanosis. Reflex apnea was present in four of the six patients (67%). In fact, one child had a complete respiratory arrest following eating. Additionally, two children had associated congenital anomalies (cardiovascular and gastrointestinal). Tracheobronchitis and episodes of upper respiratory tract infections were common, and four of the six (67%) had at least one episode.

**FINDINGS**

Lateral chest roentgenograms demonstrated tracheal compression (Figure 2) in all the cases. Although esophograms were performed in all patients, none were diagnostic for innominate artery compression. Arteriography was performed only when congenital cardiac abnormalities were either suspected or had been previously diagnosed. The tidal breathing flow volume loop was recently instituted in the workup of these patients and was used as a diagnostic test in two patients. Both exhibited marked flattening of the expiratory limb (Figure 3) compatible with obstruction distal to the larynx.
At bronchoscopy, a typical finding in all patients was that of ventral-lateral compression of the trachea at the level between its middle and lower third. The compression was pulsatile and the degree of occlusion of the lumen varied from slight to severe, causing almost complete obstruction during expiration. Diminution of the right radial pulse was elicited in all cases by elevation of the tip of the bronchoscope against the area of compression (Figure 4).

**TREATMENT**

Management of the patients was divided into medical and surgical. In those patients who presented with no reflex apnea and less than 75% narrowing of the trachea (two of six), conservative medical management consisting of humidified oxygen and observation was instituted. Surgical intervention consisting of suspension of the innominate artery was indicated in those who manifested both reflex apnea and compression greater
Fig. 2. Lateral chest roentgenograph showing tracheal narrowing (arrow) secondary to compression by the innominate artery.

than 75% (four of six). Follow-up on the patients (Table II) indicated no apneic episodes following surgical operation.

DISCUSSION

Despite reports indicating that a plain lateral chest film is the primary diagnostic tool in diagnosis of innominate artery compression,9,10 Ardito and his coworkers concluded that chest radiography may only raise the suspicion of the existence of tracheal compression by the innominate artery.11 They felt that an absolute diagnosis can best be made by endoscopic evaluation of the airway.

Abramson et al. recently introduced the tidal breathing flow volume loop as a diagnostic technique to evaluate upper airway disease in neo-
Fig. 3. Maximum and tidal flow volume loop showing marked flattening of the expiratory limb (arrow) typically seen in airway obstruction distal to the larynx.

Fig. 4. Diminution of right radial pulse after elevation of tip of the bronchoscope.
TABLE I INNOMINATE ARTERY COMPRESSION. LONG ISLAND JEWISH-HILLSIDE MEDICAL CENTER, 1978-1982

<table>
<thead>
<tr>
<th>Patient</th>
<th>Age at diagnosis (months)</th>
<th>Sex</th>
<th>Respiratory distress</th>
<th>Reflex Apnea</th>
<th>Cyanosis</th>
<th>Associated congenital anomalies</th>
</tr>
</thead>
<tbody>
<tr>
<td>1. C.M.</td>
<td>5</td>
<td>M</td>
<td>+</td>
<td>+</td>
<td>+</td>
<td>-</td>
</tr>
<tr>
<td>2. D.B.</td>
<td>10</td>
<td>F</td>
<td>+</td>
<td>-</td>
<td>-</td>
<td>+</td>
</tr>
<tr>
<td>3. J.A.</td>
<td>22</td>
<td>M</td>
<td>+</td>
<td>+</td>
<td>+</td>
<td>+</td>
</tr>
<tr>
<td>4. J.N.</td>
<td>4</td>
<td>F</td>
<td>+</td>
<td>+</td>
<td>+</td>
<td>-</td>
</tr>
<tr>
<td>5. K.M.</td>
<td>6</td>
<td>F</td>
<td>+</td>
<td>+</td>
<td>+</td>
<td>-</td>
</tr>
</tbody>
</table>

TABLE II RESULTS OF SURGICAL INTERVENTION

<table>
<thead>
<tr>
<th>Patient</th>
<th>Age at surgery (months)</th>
<th>Follow-up (months)</th>
<th>Apnea/cyanosis</th>
</tr>
</thead>
<tbody>
<tr>
<td>1. C.M.</td>
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<td>19</td>
<td>-</td>
</tr>
<tr>
<td>2. J.N.</td>
<td>4</td>
<td>42</td>
<td>-</td>
</tr>
<tr>
<td>3. M.E.</td>
<td>3</td>
<td>57</td>
<td>-</td>
</tr>
<tr>
<td>4. K.M.</td>
<td>6</td>
<td>42</td>
<td>-</td>
</tr>
</tbody>
</table>

nates and infants. This simple, noninvasive procedure demonstrates distinct and recognizable flow volume loop patterns for both intra and extrathoracic upper airway obstruction. Its use in patients suspected of innominate artery compression of the trachea is both desirable and essential since its noninvasive nature and reproducibility make it an ideal diagnostic test. Since its introduction in our institution, the diagnosis of innominate artery compression has been made preoperatively on two children presenting with reflex apnea. Subsequent endoscopy confirmed this diagnosis and determined the amount of tracheal compression.

We agree with those authors who advocate surgical suspension in those patients who present with reflex apnea and tracheal obstruction greater than 75%. In our series, four such patients were defined and none exhibited any evidence of apnea or stridor following surgery.

Despite initial doubt as to the existence of tracheal compression by the innominate artery, today it is felt that significant obstruction of respiration associated with reflex apnea is a life-threatening situation that must be identified. The tidal breathing flow volume loop offers an accurate, noninvasive way to diagnose this entity. In those patients with reflex apnea and marked tracheal compression, surgical suspension has proved efficacious in eliminating all of the preoperative symptoms.

REFERENCES


