Henryk Mazurek[1, 2], Anna Brębborowicz[3], Zbigniew Doniec[4], Andrzej Emeryk[5], Katarzyna Krenke[6], Marek Kulus[6], Beata Zielnik-Jurkiewicz[7]

[1]Department of Pneumonology and Cystic Fibrosis, Institute of Tuberculosis and Pulmonary Diseases, Rabka-Zdrój, Poland
[2]State Higher Vocational School, Nowy Sącz, Poland
[3]Department of Pneumonology, Pediatric Allergy and Clinical Immunology, Poznan University of Medical Science, Poznań, Poland
[4]Department of Pneumonology, Institute of Tuberculosis and Pulmonary Diseases, Rabka-Zdrój, Poland
[5]Department of Pulmonary Diseases and Children Rheumatology, Medical University of Lublin, Lublin, Poland
[6]Department of Pediatric Pneumonology and Allergy, Medical University of Warsaw, Warsaw, Poland
[7]Department of Otolaryngology, Children's Hospital, Warsaw, Poland

Acute subglottic laryngitis. Etiology, epidemiology, pathogenesis and clinical picture

Abstract

In about 3% of children, viral infections of the airways that develop in early childhood lead to narrowing of the laryngeal lumen in the subglottic region resulting in symptoms such as hoarseness, a barking cough, stridor, and dyspnea. These infections may eventually cause respiratory failure. The disease is often called acute subglottic laryngitis (ASL). Terms such as pseudocroup, croup syndrome, acute obstructive laryngitis and spasmodic croup are used interchangeably when referencing this disease. Although the differential diagnosis should include other rare diseases such as epiglottitis, diphtheria, fibrinous laryngitis and bacterial tracheobronchitis, the diagnosis of ASL should always be made on the basis of clinical criteria.

Key words: subglottic laryngitis, croup, laryngeal obstruction, inspiratory dyspnoea, stridor

Adv Respir Med. 2019; 87: 308–316

Definition and nomenclature

In approximately 3% of children [1, 2], viral respiratory tract infections that develop in early childhood lead to a narrowing of the larynx and include symptoms such as hoarseness, a 'barking' cough, stridor, and sometimes dyspnea. These pathological changes may eventually cause respiratory failure. In Poland, the disease is often referred to as acute subglottic laryngitis, but some different terms are used interchangeably.

The English language includes many terms for the disease such as *subglottic laryngitis, pseudocroup*, *croup*, *laryngitis*, *laryngotracheitis*, *laryngotracheobronchitis*, *laryngotracheobronchopneumonitis, viral croup* and *spasmodic croup* [3, 4]. Although these terms are sometimes used interchangeably (both in Polish and in English), they are not synonymous. The one element that they have in common is laryngitis. However, some of them also indicate the location of the lesions or their pathological background (e.g. *subglottic laryngitis*, *viral croup*). Semantic differences may lead to misunderstanding the cause of the disease and the subsequent necessary treatment (e.g. *laryngitis vs. subglottic laryngitis*).

The terms commonly used for acute laryngeal obstruction during the course of a respiratory tract infection are defined as the following (in alphabetical order):

— croup — the term traditionally designed to describe laryngeal diphtheria (ICD10: A36.2) caused by Corynebacterium diphtheriae. No cases have been reported in Poland in recent years (the result of obligatory vaccinations). In medical literature, the term is sometimes used in relation to subglottic laryngitis (duplicating the English-language term croup);

Address for correspondence: Henryk Mazurek, Clinic of Pneumology and Cystic Fibrosis, Regional Department of the Institute of Tuberculosis and Pulmonary Diseases, Rabka-Zdrój, Poland; e-mail: hmazurek@igrabka.edu.pl
DOI: 10.5603/ARM.2019.0056
Received: 14.06.2019

ISSN 2451–4934

— pseudocroup (viral croup) — similar clinical picture to the aforementioned "croup" but with a different etiology;
— spasmodic croup (spasmodic laryngitis) — a term formerly used to define recurrent laryngeal obstruction suggesting a "spastic" mechanism of obstruction. However viruses are isolated from the nasopharynx of children with a similar frequency, during the first and subsequent episodes [5], and with no improvement observed after asthma treatment [6]. Relapses require further diagnostic steps, to identify in particular coexisting central airway obstruction, gastroesophageal reflux and immunological deficits [7];
— acute obstructive laryngitis (ICD 10: J05.0 — from Polish ICD 10 translation — "croupous" laryngitis) — the "obstructive" nature of the disease is highlighted;
— acute subglottic laryngitis — the most precise term which brings attention to the dominant location of the inflammation. It refers to a specific form of acute laryngitis associated with obstruction in the place of transition between the larynx and the trachea;
— laryngitis (less common; laryngitis and tracheitis; from Latin; laryngitis acuta/diffusa — ICD 10: J04.0 / laryngotracheitis (ICD 10: J04.2) / laryngotracheobronchitis / laryngotracheobronchopneumonitis) — general terms for laryngitis possibly involving the trachea, bronchi (typical of viral infections) and/or even the lungs (which is debatable). When using these terms, there is no suggestion of laryngeal obstruction;
— croup syndrome — a term comprising a heterogeneous group of infectious forms of laryngitis of different etiology and location [4] whose common feature is the obstruction of the central airways. According to Hanicka [8], this syndrome includes pseudocroup (viral croup), diphtheria, epiglottitis, fibrinous laryngotracheobronchitis. According to Sharma [4], the syndrome includes laryngotracheitis, laryngotracheobronchitis, laryngotracheobronchopneumonitis, spasmodic croup and bacterial tracheitis. A viral infection (subglottic laryngitis) is the most common cause, but epiglottitis has the highest risk of mortality.

Summary 1.

The authors of this document agree that acute subglottic laryngitis is the most precise term to define laryngeal obstruction in the course of viral upper respiratory tract infections. Other commonly used medical terms may be considered acceptable, but acute subglottic laryngitis is preferred.

Etiology

Acute laryngeal obstruction most frequently occurs during the course of a viral respiratory tract infection in young children (~98%) [9]. The parainfluenza viruses (especially type 1; less frequently type 2 or 3) are responsible for around ½–¾ cases. The special role of parainfluenza viruses is explained by their ability to activate chlorine secretion and to inhibit sodium absorption. The cumulative effect of both these processes results in an intense edematous reaction. Acute laryngeal obstruction in the course of a viral infection can also be caused by RSV, influenza viruses, rhinoviruses, adenoviruses, enteroviruses, coronaviruses, bocaviruses and metapneumoviruses. In unvaccinated children, the condition can be observed in measles (usually with severe obstruction). Herpes simplex virus has also been associated with a severe course of the disease. Less commonly, acute laryngeal obstruction can also be observed in other bacterial respiratory infections such as epiglottitis, diphtheria and fibrinous laryngotracheobronchitis. *Mycoplasma pneumoniae* [10] is also mentioned as an etiological factor.

Summary 2.

Subglottic laryngitis most frequently occurs during the course of a viral infection caused by the parainfluenza virus.

Epidemiology

The disease is frequently reported in infants and young children in their first four years of life. It is most commonly diagnosed between 6 months and 5 years of age, peaking at 2 years of age [2]. It rarely occurs in newborns and infants < 3 months of age. This may result from the protective role of the mother's IgG antibodies. The disease affects about 3% of children and is 1.4–2 times more prevalent in boys than in girls [11]. In the United States, visits to the emergency department due to acute subglottic laryngitis constitute up to 15% of all visits that are caused by respiratory diseases [12].

The disease occurs throughout the whole year, but most commonly occurs in fall months. Some studies show a higher incidence of cases

in odd years, which can be explained by the increased number of parainfluenza infections reported every 2 years [13].

In some children (mainly boys), subglottic laryngitis is a recurrent disease. Symptoms recur during respiratory infections. This trend decreases over time, which can be partially explained by the natural effect of the airways growing in size due to the natural growing process. In a retrospective Belgian study [14], at least 1 episode of acute subglottic laryngitis was reported in 16% of children aged 5–8 years; 5% experienced relapses (at least 3 episodes). In children with recurrent episodes, predisposing factors should be taken into account. An analysis of a group of 235 children with recurrences of acute subglottic laryngitis (at least 3 times a year) showed abnormalities in bronchoscopy in 61.7% of cases, including 11.5% of cases which required surgical intervention [15]. A bronchoscopy revealed features suggestive of gastroesophageal reflux in 56% of cases. Other anomalies included tracheomalacia, subglottic narrowing of the larynx, and compression of the trachea by the brachiocephalic artery. In some children, the cause of relapse was multifactorial. Greifer *et al.* [16] found respiratory abnormalities in 53% of children with recurring croup, tracheomalacia being the most common finding. In children that experienced episodes of laryngeal obstruction during the course of viral infections where close family members were observed to present with similar symptoms, the risk of relapse is 3 to 4 times greater [17].

Epidemiologically, a relationship was found which confirmed a higher risk of bronchial asthma after the occurrence of acute subglottic laryngitis [18]. During the 5-year follow-up observation of children with acute subglotottic laryngitis, the hazard ratios for asthma were 2.13 in those aged below 5 years, and 2.22 in those between 6 and 12 years. Boys with croup had a higher risk for asthma, as well as those living in cities compared to those living in the countryside (HR = 1.78). These surveys suggest that further observation of children with recurrent subglottic laryngitis is necessary to potentially screen for asthma [18]. The possible mechanisms that explain the increased risk of asthma in children with a history of acute subglottic laryngitis include a smaller diameter of the airways (both central and peripheral), more frequent atopy, gastroesophageal reflux, bronchial hyperresponsiveness and common CD14 gene polymorphisms [19–22]. In adulthood, cases of acute subglottic laryngitis are very rare [23].

Summary 3.

The disease occurs mainly in children between 6 months and 5 years of age. Some children tend to relapse.

Pathogenesis

The predisposing factors for acute subglottic laryngitis in children are the shape and size of the larynx, a tendency for submucosal swelling (especially in the subglottic region), and airway hyperreactivity.

A viral infection of the respiratory tract is usually associated with a diffuse inflammatory reaction of the airway mucous membrane (from the nasal cavity to the bronchi), with the presence of congestion, mucosal swelling, epithelial necrosis and desquamation. The intensity of inflammation depends, among other things, on the type of pathogen as well as on individual predispositions. Symptoms of acute subglottic laryngitis are caused by edema within laryngeal mucosa, leading to narrowing of the lumen and subsequently, airflow disturbances.

Laryngeal obstruction is observed only in some children with respiratory tract infections, possibly due to anatomic predisposition (smaller airway diameter) or abnormal immune response (excessive inflammatory and edematous reaction). This is confirmed by a number of epidemiological studies including the cohort analysis from Tuscon which showed higher values of inspiratory resistance in infants who developed later episodes of stridor without wheezing [19].

The most common location of obstruction in children is the subglottic region. This is because of the specific anatomical structure of the larynx in this age group. This is the narrowest part of the airway (at the level of the cricoid cartilage). In the larynx, especially in the subglottic region, the submucosa contains significant amounts of connective tissue which can easily get swollen with clinically significant obstruction as a result of the action of inflammatory mediators. Because of its shape and structure, there is no possibility for the cricoid cartilage to dilate so swelling causes a reduction in the internal diameter. This leads to an increase in airflow resistance (both inspiratory and expiratory), which is inversely proportional to the fourth power of the radius of the laryngeal diameter. In the case of turbulent flow, this resistance can rise to the fifth power.

Respiratory effort during ventilation significantly increases (inspiratory pleural cavity pressure falls to about 40–50 cm H_2O, instead of

to approximately 10 cm H_2O). To a lesser extent, the expiratory pressure also increases (up to 10 cm H_2O).

The increased amplitude of inspiratory pressure (due to the activation of accessory inspiratory muscles) leads to the collapse of supple extra-thoracic structures with subsequent increase in both inspiratory resistance and stridor. Extremely severe obstruction can lead to the asynchronous movement of the chest wall and abdomen (paradoxical breathing), fatigue, decreased airflow (paradoxically, stridor may be diminished) and finally, to respiratory failure.

In mild to severe acute subglottic laryngitis, air flow resistance is increased, especially during inspiration (about fourfold increase). To a lesser extent, airflow resistance is also increased during exhalation (about twofold). Work of breathing increases threefold [24].

Minute ventilation is maintained by an increase in the respiratory rate and a decrease in tidal volume (TV). The time of inspiration and expiration shortens, but the proportion of these phases with minor obstruction remains unchanged. With greater obstruction, the inspiratory phase is prolonged [24].

Because of the reduction in airway diameter, the linear velocity of the airflow in the larynx increases and the airflow becomes turbulent. A significant rise in the pressure amplitude usually allows for maintaining normal inspiratory flow, although the expiratory flow is reduced [especially peak expiratory flow (PEF) — by around 1/5]. The flow ratio is reversed; in healthy children, the mid-expiratory flow is higher than the mid-inspiratory flow. However, in laryngeal obstruction, the flow during inspiration is higher. The ratio of the time necessary to achieve peak flow during exhalation to the time of exhalation increases (delay in the exhalation peak) [24].

Stridor is one of the key symptoms of laryngeal obstruction. This term refers to the harsh, often high-tone sound created by the rapid, turbulent flow of air through the narrowed large airways in the extra-thoracic segment. It is heard mainly during inspiration and is generated by the vibrating walls of the central part of the respiratory tract [25]. Inspiratory stridor originates in the glottis (or above), while expiratory stridor comes from the structures located below that level (trachea, bronchi). The presence of stridor depends not only on the degree of narrowing in the central airways, but also on the flows and pressures generated.

For stridor evaluation it is important to analyze the following: breathing phase, sound intensity, pitch, and the conditions of its generation (in particular, its presence at rest).

Summary 4.

The anatomical and pathophysiological specificity of the larynx in early childhood make this group more predisposed to acute subglottic laryngitis. The main consequence of the inflammatory reaction within the larynx is inspiratory airflow disturbance with varying degrees of severity.

The clinical picture

The symptoms of the disease appear suddenly, mostly in previously healthy children and usually at night a few hours after falling asleep. The most characteristic symptom is a dry, barking cough ("seal-like") which is most often seen in young children. The patient's voice is usually clear, although it is sometimes accompanied by hoarseness and, on rare occasions, aphonia. As the narrowing of the larynx continues, other symptoms appear, including stridor of variable intensity and inspiratory dyspnea. When there is further increase in obstruction, stridor can also be heard during expiration. Anxiety and crying exacerbate the symptoms and make the stridor louder.

These symptoms often disappear spontaneously after a few hours. Sometimes, they can be severe.

Usually 1 to 2 days before the onset of laryngitis there are mild symptoms such as serous discharge from the nose, mildly elevated body temperature (rarely fever), sore throat, anxiety, a loss of appetite and/or malaise.

In severe cases, the body temperature can be elevated and this is accompanied by anxiety, rapid breathing, shortness of breath and dyspnea. Increased work of the accessory respiratory muscles (supra- and subclavicular, intercostal, abdominal muscles and diaphragm) is also common. The nasal flaring intensifies. Suprasternal and chest wall retractions can be present.

In its most severe forms, symptoms include: anxiety, agitation or confusion, respiratory failure, cyanotic or pale skin (around nose and mouth), apnea, loss of consciousness and cardiovascular failure.

On auscultation, the breath sounds are usually slightly exacerbated, and no wheezing is observed on expiration. Laryngoscopy reveals swelling of the laryngeal mucosa. There are no problems with swallowing and drooling is not noted. There is also no deterioration in the child's general condition

(which illustrates the common finding that the "child looks better then you can hear").

Summary 5.

The symptoms of the disease include stridor and a dry, barking cough beginning suddenly, usually at night. Severe laryngitis leading to respiratory failure is rare.

Severity assessment

There are no universally accepted classification methods for severity assessment of laryngeal obstruction. Among many, only the 17-point Westley croup score has been validated (Table 1). It is mainly used for scientific research and rarely in clinical practice.

A quick assessment may be facilitated by simpler, albeit non-validated scales, such as the Geelhoed scale (Table 2) which is based on an assessment of stridor intensity and retraction of the chest wall [27].

In severity assessment, it is essential to evaluate the degree of airway obstruction, respiratory problems, and the dynamics of the disease. It is also important to take into consideration the course of possible previous episodes.

In clinical practice, the assessment of severity is based on symptoms (medical history + an examination of the child) and includes:

— the general condition — the most important exponent of severity. The important indicators are the child's activity and the ability to take in food and fluids;
— the degree of respiratory problems (the severity of stridor, the presence of chest retractions, presence of paradoxical breathing, and auscultatory symptoms);
— other — skin colour changes (cyanosis or pallor of the skin), gas exchange disturbances (SpO2, blood gases).

In **mild** obstruction (Westley croup score 0–2 points), the child is in good condition, is active and is willing/capable to take in fluids and food. The symptoms include a mild, mainly "barking cough" with hoarseness. There is no stridor at rest, which can be observed only during hyperventilation (e.g. crying). Retractions of the chest are either non-existent or very minor [28, 29].

Moderate (medium) obstruction in acute subglottic laryngitis (Westley croup score 3–5 points) is characterized by an intensive "barking" cough, inspiratory stridor at rest, chest wall retractions and tachycardia. When the child is not agitated, the condition is normal.

In **severe** obstruction (Westley croup score 6–11 points), the cough is intensive, with loud and often biphasic stridor (inspiratory — expiratory). There are retractions of the chest walls. The main symptom of severe obstruction is an

Table 1. Westley croup score [26]

Score (range)	Stridor (0–2)	Retractions (0–3)	Air entry (0–2)	Cyanosis (0–5)	Level of consciousness (0–5)
0	None	None	Normal	None	Normal (including sleep)
1	When agitated	Mild	Decreased		
2	At rest	Moderate	Markedly decreased		
3		Severe			
4				Cyanosis with agitation	
5				Cyanosis at rest	Disoriented

≤ 2 — mild; 3–7 — moderate; 8–11 — severe; ≥ 12 — impending respiratory failure

Table 2. Geelhoed score [27]

Score	Stridor	Retractions
0	None	None
1	Only on crying, exertion	Only on crying, exertion
2	At rest	At rest
3	Severe (biphasic)	Severe (biphasic)

1–2 — mild; 3–4 — moderate; 5–6 — severe

increased duration of the expiratory phase and active action of the abdominal muscles. The child will present with anxiety and agitation.

Life-threatening laryngeal obstruction (impending respiratory failure, Westley croup score 12–17 points) is characterized by tachycardia with cough of variable intensity. The cough, similarly to stridor, may decrease at the terminal stage of respiratory failure due to respiratory muscle fatigue and reduced airflow. Sternal retraction may be observed. The child will present as fatigued and exhausted. This is usually the result of apathy which follows a period of anxiety. The child will also present with skin color changes (cyanosis, pallor) and a depressed level of consciousness.

Summary 6.

Besides clinical symptoms, there is no universally applicable standard in order to assess the severity of laryngeal obstruction. The Westley croup score may be useful for further management.

Laboratory studies and imaging

Further laboratory studies and imaging are not required in typical cases. They are justified only in cases where another diagnosis is suspected.

Blood tests may show an increase in some inflammatory markers such as ESR. Lymphocytosis in a peripheral blood smear may also be present. An increased CRP has been observed in very rare cases of acute subglottic laryngitis in adults [23].

Radiographic imaging is not routinely recommended. It may be useful if the location of obstruction is in question in order to differentiate the disease from epiglottitis. In about one-half of children suffering from the disease, a radiological examination of the larynx performed in the anterior-posterior projection shows a marked narrowing in the subglottic region (the tower sign = the steeple sign = the pencil sign = the wine bottle sign). Dynamic investigations show ~60% narrowing of the anteroposterior dimension of the extra-thoracic part of the trachea during inspiration [24]. A radiological image can capture narrowing in the subglottic region, but it does not show the severity of the obstruction [30]. In some cases (differentiation with epiglottitis), a lateral projection (thumb sign) is indicated. This confirms a diagnosis of epiglottitis.

A small decrease in SpO_2 and pO_2 is often observed. However, it is mainly an indicator of lung function and probably results from the simultaneous involvement of the lower respiratory tract (venous blood shunt). Initially, mild hypoxemia with respiratory alkalosis can be observed (due to hypocapnia). A continued increase in obstruction may lead to a subsequent decrease in pO_2 and in the possibility of CO_2 accumulation (very rare, only in the most severe cases). Therefore, arterial blood gases are not useful in the assessment of the function of the upper respiratory tract in mild subglottic laryngitis [24].

Some respiratory function tests can record movements of the chest and abdomen. They can also measure tidal volume as well as pressures in the lower part of the esophagus. Respiratory flows can be measured via spirometry in older children. However, in younger patients, a tightly fitting mask with a pneumotachometer is required. Measurements of flows and pressures in the lower part of the esophagus (corresponding to the pressure in the pleural cavity) allow for the calculation of airflow resistance [24]. However, none of these tests have been applied in routine practice.

Antigen tests or viral tests are not necessary for most children with laryngitis.

Summary 7.

Laboratory tests or other investigations are not required in the course of typical acute subglottic laryngitis. They can be justified only in the most severe cases or when the diagnosis is in question.

Natural course and prognosis

In most of the children, the disease is mild, and symptoms subside even without intervention. However, the severity of laryngeal obstruction and the child's condition can change rapidly (risk of sudden deterioration). The symptoms usually worsen at night, which can be explained by the cyclic course of physiological processes (especially changes in cortisol levels).

Canadian studies showed that the mild form of the disease affects 85% of children, while severe symptoms are reported in less than 1%. As a consequence, the majority of children can be treated on an outpatient basis. In 60% of children, laryngeal obstruction subsides within 48 hours. In over 75% of patients, the obstruction subsides within 72 hours. Only a few cases have been reported where children had symptoms persisting for a week [13, 29].

It is estimated that less than 5% of children need admission to a hospital (from 1.5% to 31%

of children seen at an outpatient clinic). Only 1–3% of admitted patients require intubation [12, 13, 19]. The length of hospital stay is longer for younger children (infants) and boys [31]. Deaths due to respiratory failure are rare and observed in less than < 0.5% of all intubated children (~1/30,000 cases). The cause is usually rapidly progressing laryngeal edema.

In children with recurrent laryngitis, the frequency and severity of symptoms decrease with age and they usually subside between 6–12 years of age (as a result of airway diameter increase and maturation of the immune system).

Summary 8.

In most of the children, the course of acute subglottic laryngitis is mild and usually resolves within 3 days. However, about 5% of children need hospital admission and 1–3% of them require intubation.

Complications

Complications are rare. Most commonly, viral infection can spread to other parts of the respiratory system (the bronchi, bronchioles and the middle ear). Bacterial superinfection, pneumonia, bacterial tracheitis and complications following a tracheostomy can be observed [13, 29].

Diagnosis

A diagnosis is established on the basis of clinical symptoms. In a typical course of disease, no further laboratory tests are necessary.

Recurrent episodes, atypical course of disease, rising suspicion of predisposing causes (i.e. a narrowing of the larynx or trachea), and gastroesophageal reflux are just a few examples of situations that indicate a need for an endoscopic examination. A direct laryngeal examination should assess the appearance of the epiglottis, the laryngeal vestibule, the subglottic region, and the mobility/colour of the vocal folds. During the acute phase of the disease, a diagnosis of subglottic laryngitis can be confirmed by indirect or direct laryngoscopy which shows subglottic edema.

Differential diagnosis

Acute subglottic laryngitis requires differentiation with conditions that demand a different course of management (Table 3), especially in the case of epiglottitis [32]. Detailed differential diagnosis is especially needed for young children (< 6 months of age) or those with recurrent laryngeal obstruction (≥ 4×/year), particularly those above 6 years of age. Other indications include a very severe course of the disease, poor management outcome, the need for hospital admission, an unusual presentation (e.g. chronic stridor, increase of laryngeal obstruction in special body position), a history of laryngeal trauma, suspicion of congenital malformations and/or the presence of neurological disorders [33].

Table 3. Differential diagnosis of laryngeal obstruction [25, 32]

Course	
Acute	Subglottic laryngitis
	Aspiration of a foreign body
	Epiglottitis
	Quincke edema (e.g. anaphylaxis)
	Laryngeal diphtheria
	Bacterial tracheitis
	Hypocalcaemia
Subacute	Abscess:
	— retro-pharyngeal
	— peri-tonsillar
Chronic	Post-intubation stenosis
	Laryngeal papillomas
	Congenital changes:
	— laryngo/tracheomalacia
	— paralysis of the vocal cords
	— laryngeal haemangioma
	— vascular ring
	— cysts
	— laryngeal cleft
	— laryngeal tumours
	— gastroesophageal reflux

An unclear medical history given by parents (without current symptoms of dyspnea) may require differentiation with asthma.

Acute epiglottitis (*acute epiglottitis = supraglottitis = laryngitis supraglottica = bacterial croup*; ICD10: J05.1) in the era of vaccination is one of the less common forms of laryngitis. *Haemophilus influenzae* type b (Hib) capsular strain is known to be the main etiological factor. Less often, other bacteria (such as *Streptococcus pneumoniae* or *Streptococcus pyogenes*) and viruses are responsible for this disorder. The bacterial inflammatory process usually affects the entire supraglottic region, including the epiglottic mucosa and aryepiglottic folds. The course of this life-threatening condition is much more severe than in subglottic laryngitis. The risk of rapid, complete airway obstruction with associated bac-

teraemia leads to the conclusion that this disease requires different management.

The disease is mainly seen in children aged 2–10 years, peaking in the third to fourth year of life (82%). It should be noted that recently there has been an increase in the incidence of infections in adults [34–39]. Morbidity may be associated with reduced levels of immune antibodies, especially in the latter case. Sometimes it can be triggered by local damage of the mucosa caused by a food bite with sharp edges that initiates bacterial penetration. Epiglottitis may occur in patients with neoplastic diseases, with *Streptococcus pneumoniae* or *Candida albicans* as the most common etiological factors [34, 38, 40–42].

Epiglottitis has an abrupt onset with a rapid and severe course. It develops within a few hours, leading to respiratory failure. The symptoms usually start suddenly without prodromal "flu-like" symptoms. Severe sore throat and pain during swallowing results in drooling and difficulties in swallowing food and fluids. Rapidly progressing inspiratory-expiratory dyspnea forces patients to take a forward-leaning sitting up position. Anxiety and general weakness are also commonly noted. Because of bacteraemia, the child is usually toxic, febrile and in poor general condition, with sudden changes in body temperature. A "barking" cough is not a typical symptom. Moist stridor with low tones predominates. There is also typically a muffled voice ("hot potato voice"), followed by aphonia.

Because of impending airway obstruction, the child's throat should not be examined in order to avoid an anxiety attack. It can trigger a complete blockage of airways due to swelling of the epiglottis. Although leukocyte count and ESR are often increased, blood sampling with blood culture should also be discouraged to avoid increasing the child's anxiety and should be delayed until the airway is secure. Lateral X-ray of the larynx may be helpful. It shows a swollen, enlarged epiglottis (the thumb sign). It is important to note that sometimes, the image can be normal. There are no abnormalities observed in the subglottic area.

An endoscopic examination (carried out only in exceptional cases in a controlled environment like an operating room or ICU) reveals the thickened, swollen, erythematous epiglottis and aryepiglottic folds, as well as narrowing of the laryngeal vestibule. Because of the risk of total airway obstruction during examination, materials should be prepared for intubation (trans-nasal) or for a tracheotomy.

Given the risk of complete laryngeal obstruction and septic course of this life-threatening condition, a child with suspicion of acute epiglottis requires hospitalization (with access to ICU). Because many patients require immediate intubation, initial treatment should be carried out in an intensive care unit. In many centers, elective intubation is preferred [42–44]. Broad-spectrum antibiotics (mainly third generation cephalosporin i.e. cefotaxime or ceftriaxone) are used either alone or in combination with a second antibiotic (i.e. clindamycin or vancomycin) and are administered intravenously [45]. The first dose should be given as soon as possible, preferably even before the patient is transferred to the hospital.

Summary 9.

Acute subglottic laryngitis is diagnosed on the basis of clinical symptoms. Unspecific symptoms and relapses indicate the need for a detailed differential diagnosis.

Acknowledgments

We would like to thank Professor Bolesław Samoliński for constructive comments.

Conflict of interest

None declared.

References:

1. Rosychuk RJ, Klassen TP, Metes D, et al. Croup presentations to emergency departments in Alberta, Canada: a large population-based study. Pediatr Pulmonol. 2010; 45(1): 83–91, doi: 10.1002/ppul.21162, indexed in Pubmed: 19953656.
2. Asher M, Grant C. Infections of the upper respiratory tract. [in:] Pediatric Respiratory Medicine. Taussig LM, Landau LI, LeSouef PN et al. (ed). Mosby, Philadelphia 2008 .
3. Russell K, Liang Y, O'Gorman K, et al. Glucocorticoids for croup. Cochrane Database of Systematic Reviews. 2011, doi: 10.1002/14651858.cd001955.pub3.
4. Sharma BS, Shekhawat DS, Sharma P, et al. Acute respiratory distress in children: croup and acute asthma. Indian J Pediatr. 2015; 82(7): 629–636, doi: 10.1007/s12098-014-1559-4, indexed in Pubmed: 25257964.
5. Wall SR, Wat D, Spiller OB, et al. The viral aetiology of croup and recurrent croup. Arch Dis Child. 2009; 94(5): 359–360, doi: 10.1136/adc.2008.142984, indexed in Pubmed: 18801765.
6. Kwong K, Hoa M, Coticchia JM. Recurrent croup presentation, diagnosis, and management. Am J Otolaryngol. 2007; 28(6): 401–407, doi: 10.1016/j.amjoto.2006.11.013, indexed in Pubmed: 17980773.
7. Pfleger A, Eber E. Assessment and causes of stridor. Paediatr Respir Rev. 2016; 18: 64–72, doi: 10.1016/j.prrv.2015.10.003, indexed in Pubmed: 26707546.
8. Hanicka M. Ostre choroby układu oddechowego [in:] Ostre i przewlekłe choroby układu oddechowego u dzieci. Rudnik J, Hanicka M (ed.). PZWL, Warszawa 1978: 97–151.
9. Primhak R. Evaluation and management of upper airway obstruction. Paediatrics and Child Health. 2013; 23(7): 301–306, doi: 10.1016/j.paed.2012.11.008.

10. Rihkanen H, Rönkkö E, Nieminen T, et al. Respiratory viruses in laryngeal croup of young children. J Pediatr. 2008; 152(5): 661–665, doi: 10.1016/j.jpeds.2007.10.043, indexed in Pubmed: 18410770.
11. Denny FW, Murphy TF, Clyde WA, et al. Croup: an 11-year study in a pediatric practice. Pediatrics. 1983; 71(6): 871–876, indexed in Pubmed: 6304611.
12. Zoorob R, Sidani M, Murray J. Croup: an overview. Am Fam Physician. 2011; 83(9): 1067–1073, indexed in Pubmed: 21534520.
13. Bjornson C, Johnson D. Croup. The Lancet. 2008; 371(9609): 329–339, doi: 10.1016/s0140-6736(08)60170-1.
14. Van Bever HP, Wieringa MH, Weyler JJ, et al. Croup and recurrent croup: their association with asthma and allergy. An epidemiological study on 5-8-year-old children. Eur J Pediatr. 1999; 158(3): 253–257, doi: 10.1007/s004310051062, indexed in Pubmed: 10094451.
15. Duval M, Tarasidis G, Grimmer JF, et al. Role of operative airway evaluation in children with recurrent croup: a retrospective cohort study. Clin Otolaryngol. 2015; 40(3): 227–233, doi: 10.1111/coa.12353, indexed in Pubmed: 25409938.
16. Greifer M, Santiago MT, Tsirilakis K, et al. Pediatric patients with chronic cough and recurrent croup: the case for a multidisciplinary approach. Int J Pediatr Otorhinolaryngol. 2015; 79(5): 749–752, doi: 10.1016/j.ijporl.2015.03.007, indexed in Pubmed: 25818348.
17. Pruikkonen H, Dunder T, Renko M, et al. Risk factors for croup in children with recurrent respiratory infections: a case-control study. Paediatr Perinat Epidemiol. 2009; 23(2): 153–159, doi: 10.1111/j.1365-3016.2008.00986.x, indexed in Pubmed: 19159401.
18. Lin SC, Lin HW, Chiang BL. Association of croup with asthma in children: A cohort study. Medicine (Baltimore). 2017; 96(35): e7667, doi: 10.1097/MD.0000000000007667, indexed in Pubmed: 28858086.
19. Castro-Rodríguez JA, Holberg CJ, Morgan WJ, et al. Relation of two different subtypes of croup before age three to wheezing, atopy, and pulmonary function during childhood: a prospective study. Pediatrics. 2001; 107(3): 512–518, doi: 10.1542/peds.107.3.512, indexed in Pubmed: 11230591.
20. Arslan Z, Cipe FE, Ozmen S, et al. Evaluation of allergic sensitization and gastroesophageal reflux disease in children with recurrent croup. Pediatr Int. 2009; 51(5): 661–665, doi: 10.1111/j.1442-200X.2009.02859.x, indexed in Pubmed: 19419517.
21. Modaresi M, Pourvali A, Azizi G, et al. Association of childhood croup and increased incidence of airway hyperreactivity in adulthood. J Educ Health Promot. 2018; 7: 97, doi: 10.4103/jehp.jehp_138_17, indexed in Pubmed: 30159343.
22. Rennie DC, Karunanayake CP, Chen Y, et al. CD14 gene variants and their importance for childhood croup, atopy, and asthma. Dis Markers. 2013; 35(6): 765–771, doi: 10.1155/2013/434920, indexed in Pubmed: 24347797.
23. Tachibana T, Orita Y, Makino T, et al. Prognostic factors and importance of recognition of adult croup. Acta Otolaryngol. 2018; 138(6): 579–583, doi: 10.1080/00016489.2017.1422140, indexed in Pubmed: 29310492.
24. Argent AC, Newth CJL, Klein M. The mechanics of breathing in children with acute severe croup. Intensive Care Med. 2008; 34(2): 324–332, doi: 10.1007/s00134-007-0910-x, indexed in Pubmed: 18008061.
25. Boudewyns An, Claes J, Van de Heyning P. Clinical practice: an approach to stridor in infants and children. Eur J Pediatr. 2010; 169(2): 135–141, doi: 10.1007/s00431-009-1044-7, indexed in Pubmed: 19763619.
26. Westley CR, Cotton EK, Brooks JG. Nebulized racemic epinephrine by IPPB for the treatment of croup: a double-blind study. Am J Dis Child. 1978; 132(5): 484–487, doi: 10.1001/archpedi.1978.02120300044008, indexed in Pubmed: 347921.
27. Geelhoed GC, Macdonald WB. Oral and inhaled steroids in croup: a randomized, placebo-controlled trial. Pediatr Pulmonol. 1995; 20(6): 355–361, doi: 10.1002/ppul.1950200604, indexed in Pubmed: 8649914.
28. Bjornson C, Russell K, Vandermeer B, et al. Nebulized epinephrine for croup in children. Cochrane Database Syst Rev. 2013(10): CD006619, doi: 10.1002/14651858.CD006619.pub3, indexed in Pubmed: 24114291.
29. Johnson D. Croup. BMJ Clin Evid. 2009; 3(321): 1–41.
30. Hammer J. Acquired upper airway obstruction. Paediatr Respir Rev. 2004; 5(1): 25–33, doi: 10.1016/j.prrv.2003.09.007, indexed in Pubmed: 15222951.
31. Godden CW, Campbell MJ, Hussey M, et al. Double blind placebo controlled trial of nebulised budesonide for croup. Arch Dis Child. 1997; 76(2): 155–158, doi: 10.1136/adc.76.2.155, indexed in Pubmed: 9068309.
32. Mazurek H. Ostre zapalenia krtani [in:] Zakażenia układu oddechowego u dzieci. Mazurek H. (ed.). Medical Tribun, Warszawa. 2014: 86–109.
33. Mehrotra S, Kilgar J, Lim R. Case 3: Stridor and cough in a young child. Paediatr Child Health. 2014; 19(10): 523–525, doi: 10.1093/pch/19.10.523, indexed in Pubmed: 25587227.
34. Chroboczek T, Cour M, Hernu R, et al. Long-term outcome of critically ill adult patients with acute epiglottitis. PLoS One. 2015; 10(5): e0125736, doi: 10.1371/journal.pone.0125736, indexed in Pubmed: 25945804.
35. Shah RK, Stocks C. Epiglottitis in the United States: national trends, variances, prognosis, and management. Laryngoscope. 2010; 120(6): 1256–1262, doi: 10.1002/lary.20921, indexed in Pubmed: 20513048.
36. Guardiani E, Bliss M, Harley E. Supraglottitis in the era following widespread immunization against Haemophilus influenzae type B: evolving principles in diagnosis and management. Laryngoscope. 2010; 120(11): 2183–2188, doi: 10.1002/lary.21083, indexed in Pubmed: 20925091.
37. Guldfred LA, Lyhne D, Becker BC. Acute epiglottitis: epidemiology, clinical presentation, management and outcome. J Laryngol Otol. 2008; 122(8): 818–823, doi: 10.1017/S0022215107000473, indexed in Pubmed: 17892608.
38. Mathoera RB, Wever PC, van Dorsten FRC, et al. Epiglottitis in the adult patient. Neth J Med. 2008; 66(9): 373–377, indexed in Pubmed: 18931398.
39. Katori H, Tsukuda M. Acute epiglottitis: analysis of factors associated with airway intervention. J Laryngol Otol. 2005; 119(12): 967–972, doi: 10.1258/002221505775010823, indexed in Pubmed: 16354360.
40. Chen C, Natarajan M, Bianchi D, et al. Acute epiglottitis in the immunocompromised host: case report and review of the literature. Open Forum Infect Dis. 2018; 5(3): ofy038, doi: 10.1093/ofid/ofy038, indexed in Pubmed: 29564363.
41. Isakson M, Hugosson S. Acute epiglottitis: epidemiology and Streptococcus pneumoniae serotype distribution in adults. J Laryngol Otol. 2011; 125(4): 390–393, doi: 10.1017/S0022215110002446, indexed in Pubmed: 21106138.
42. Glynn F, Fenton JE. Diagnosis and management of supraglottitis (epiglottitis). Curr Infect Dis Rep. 2008; 10(3): 200–204, indexed in Pubmed: 18510881.
43. Roosevelt G. Acute inflammatory upper airway obstruction (croup, epiglottitis, laryngitis, and bacterial tracheitis). Nelson Textbook of Pediatrics. 2011: 1445–1450.e1, doi: 10.1016/b978-1-4377-0755-7.00377-8.
44. Orhan İ, Aydın S, Karlıdağ T. Infectious and noninfectious causes of epiglottitis in adults, review of 24 patients. Turk Arch Otorhinolaryngol. 2015; 53(1): 10–14, doi: 10.5152/tao.2015.718, indexed in Pubmed: 29391970.
45. Dzierżanowska D. (ed.) Przewodnik antybiotykoterapii. 21st edition. Alfa-Medica Press, Bielsko-Biała 2017.