CHAPTER 7

AIRWAY AND NERVOUS ANOMALIES ASSOCIATED WITH PATHOLOGIC SOUND PRODUCTION

IN DEALING with the individual diseases the sequence given in the Summary (Table 9-1)—proceeding from the nose down to the deeper airways—is followed. The neurologic conditions and some chromosomal anomalies accompanied by pathological cries are discussed in connection with the diseases of the larynx, whereas diseases associated with pharyngeal stridor are dealt with in the sections on nasal, oral, and pharyngeal diseases. The most important clinical features of the single diseases are also described briefly, to the extent required for understanding the pathomechanism of the acoustically analyzed sound phenomena. The new diagnostic (imaging, neurologic, and acoustic) methods, as well as up-to-date therapeutic concepts and surgical procedures are discussed.

Disorders Affecting the Nose, Oral Cavity, Pharynx, and Ears

Choanal Atresia

This condition is due to the congenital bony or membranous closure of the posterior nasal openings. In about one-third of the cases the obstruction is bilateral. The condition occurs in 1 out of every 7,000 to 8,000 live births (Dunham & Miller, 1992), but also may be part of some syndromes.

Unobstructed nasal breathing is of vital importance in infancy. Owing to the high position of the larynx in neonates, the epiglottis reaches almost to the level of the soft palate and oral breathing is inhibited by the high-lying root of the tongue. Thus, air can pass unimpeded toward deeper structures only through the nose: newborns until about the age of three to four months are obligate nasal breathers. When there is nasal obstruction, especially when it is complete and bilateral, severe respiratory disorders may arise.

The symptoms in bilateral cases include difficult breathing, increasing dyspnea during sucking, frequent episodes of aspiration, cyclic cyanosis, signs of intermittent anoxia, as well as a constant, marked stridor (Csermely & Hirschberg, 1967; Lellei & Kallay, 1974). The characteristic sound is usually more that of *stertor* than stridor (Benson, Baredes, Schwartz,

Kumar, & Clark, 2006). The unilateral atresia may remain unnoticed in a longer time: although symptoms (dense purulent nasal discharge and impeded nasal breathing on one side) exist, the parents and sometimes also the physicians do not attach importance enough to these complaints.

The stridor is typically pharyngeal. It is a noise without "harmonic" elements. It is produced between the tongue and the palate as a result of insufficient, spastic inspirations interrupting the long periods of apnea. It is mostly an interrupted, sawing, or snoring stridor. According to Myer and Cotton (1988) this is a snorting type of respiration. As the outflow of air is also obstructed, the stridor may occur during expiration as well (Figure 7-1). The cry of these infants is acoustically a clear sound, but in cases with severe obstruction the voice become hyponasal (Hirschberg et al., 2006).

KEY POINT

Choanal atresia is the congenital closure of the posterior nasal openings, it may be bony or membranous, uni- or bilateral. Unilateral atresia hardly causes any symptoms, except nasal discharge, the bilateral case presents with severe respiratory problems: dyspnea, cyanosis, and noisy pharyngeal stridor. The anomaly can be operated on transnasal or transpalatinal; in the bilateral case surgery is urgent.

Based on observation of a pharyngeal stridor the diagnosis can be easily established: if the patency of the nasal meatus cannot be demonstrated using a Politzer bag and if the catheter inserted into the

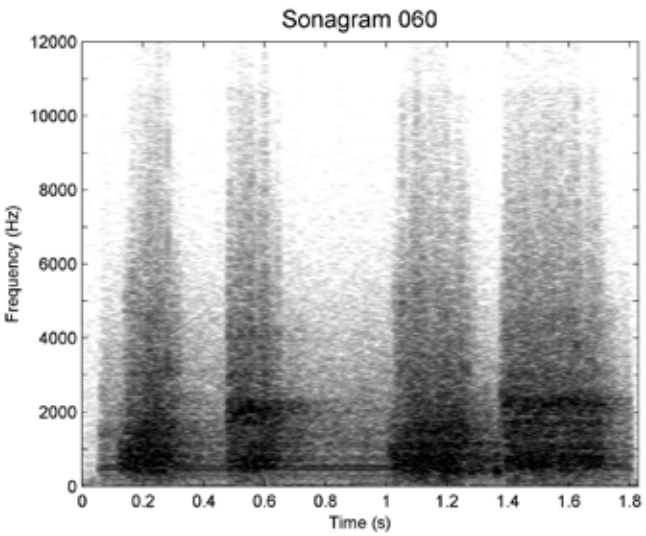
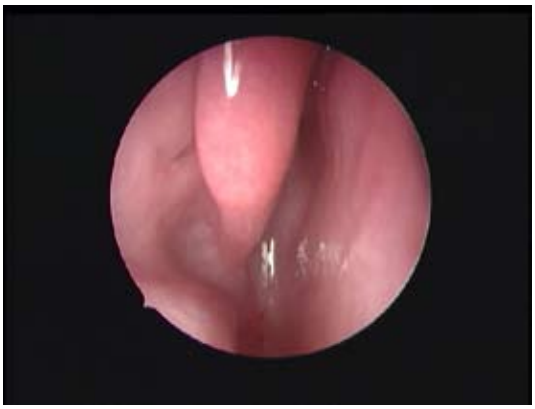


FIGURE 7-1. Bilateral choanal atresia. Infant aged 6 weeks. *History:* Frequent episodes of dyspnea; stridor which increases during feeding; recurrent attacks of cyanosis. *Acoustic finding:* Snoring pharyngeal stridor. *Sonagram No. 60:* Expiration, inspiration, expiration, and inspiration; most characteristic part between 1.2 and 1.5 s. Hardly divided noise blocks; the one around 2000 to 2500 Hz is dominant. Further characteristic components around 3000 and 5000 Hz. The slightly high-pitched and snoring character of the sound can be explained partly by the dominance of the noise components over 2000 Hz up to 11000 Hz and partly by the fundamental vibration, approximately at 500 Hz.

nasal opening does not appear in the pharynx. Some decades ago nasography was performed to furnish additional information in these cases; today the endoscopy is the preferred diagnostic tool (Figures 7-2A, B, and C), and with axial CT the thickness and extent of bone in the atretic plate may be determined (Figures 7-3A and B).

Some newborns with bilateral malformation require early intubation. *The definitive management is surgery.* The choanal atresia can be operated on transnasal or transpalatinal, depending on the extension of the obstruction and on the experience of the surgeon. Today the transnasal endoscopic atresia-solution is preferred by most authors; it is especially the best choice for thin membranous atresias in older children. The various surgical managements are summarized by Koltai (1994). After the operation nasal stents may be positioned and are usually removed 6 to 10 weeks after repair. Some authors (Katona et al., 1994) recommend to leave the child without stent after surgery; in this case local mitomycin C may be applied against cicatrization. Yaniv, Hadar, Shvero, Stern, and Raveh (2007) published a new endoscopic transnasal technique using a mucoperichondrial flap developed from the nasal septum: this is a safe and rapid procedure even in very young children, with no complication and a success rate of 91%. Cedin, Fujita, and Cruz (2006) analyzed the long-term results of a new stentless endoscopic transseptal surgical technique in 10 cases. This approach allowed resection of the posterior portion of the vomer, atretic plate, and part of the medial pterygoid process. The technique proved to be a satisfactory and



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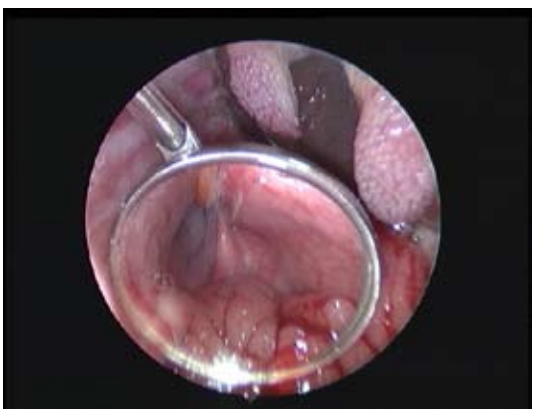

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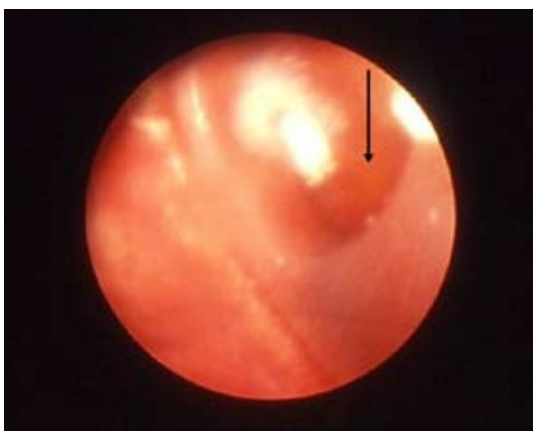

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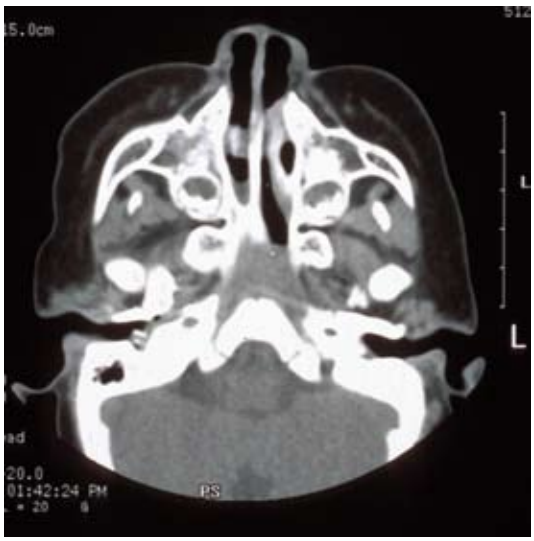
FIGURE 7-2. A and B. Right unilateral choanal atresia in a 1-year-old infant. C. The arrow shows the massive fibrous plate which closes entirely the choana. The white tissue is collected pus (endoscopic pictures).



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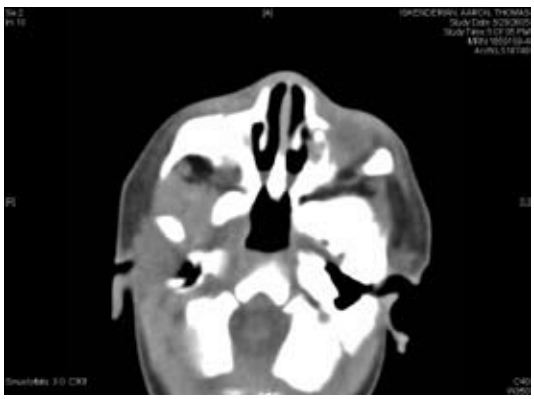

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FIGURE 7-3. A. Right unilateral choanal atresia; B. bilateral choanal atresia on CT.

safe one-step surgery using neither stents nor nasal packing.

Timing recommendations for repair of unilateral atresia vary in the literature: surgery at around 2 to 3 years of age is recommended (Jacobs, 1996). Bilateral atresia should be repaired shortly after birth. In other words: lack of contraindication means the indication of surgery in neonates (Csermely & Hirschberg, 1967).

Case Study

Bilateral choanal atresia in an infant with typical Apert syndrome (Figures 7-4A, B, and C). Immediately after birth intubation was performed because of severe respiratory distress and loud pharyngeal type stridor due to bilateral choanal atresia. At age 2 months transpalatinal surgery was performed, placing a stent (Madarász Children's Hospital, Pediatric ENT Department, Budapest). At age 5 months cranioplasty of the posterior skull; half a year later reconstruction of the anterior skull, then implanting of a shunt, later shunt-revision (Neurosurgical Institute, Budapest). At age 1 year surgery for syndactylia was performed (Orthopedic Department of St. John Hospital, Budapest). At age 2½ herniotomia scrotalis bilateralis. Half a year later widening of both choana apertures (Bethesda Children's Hospital, Budapest). At age 3 years syndactylia operation repeated (Clinic for Accident and Hand Surgery, Pécs). Half a year later surgery for hernia inguinalis recidivans and tumor Schlofferi (Madarász Children's Hospital, Surgical Department). At age 4 years grommet insertion, later hydrocele operation in Heim Pál Children's Hospital, Budapest. Repeat metal grommet insertion and bronchus lavage in ENT Clinic of Semmelweis University, Budapest. At age of 5½ years: extra- and intraoral (Le Fort III) osteotomia of the facial skull, insertion of bilateral craniofacial distractors, and dental splint of the maxilla and mandibula performed (Klinik für Mund- und Kiefer-Gesichtschirurgie, Münster). Present status: the child is 7 years old, motoric, mental and speech development is acceptable, there are no neurologic center symptoms; he

goes in school. His marks are excellent. Audiometry: 40 dB conductive hearing loss in both side, the tympanogram is C1 type. Nose breathing impeded. Nasendoscopy: severe septum deviation, both choana are obstructed with massive fibrous tissue. Management: grommet insertion, later reoperation of the choanal atresia, considering orthodontic treatment (Budai Children's Hospital, Budapest).

KEY POINT

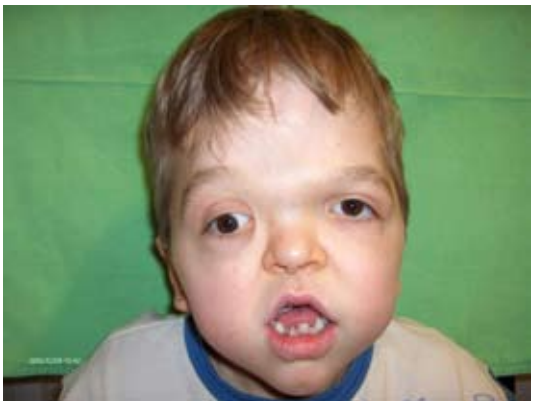
The most common cause of pharyngeal stridor is the backward displaced (and sometimes enlarged) tongue obstructing the pharynx. This condition occurs most frequently in the cases of micrognathia, in Robin sequence, in de Lange and Hurler's syndroms, as well as in some children with mental deficiency.

Micrognathia

This anomaly may occur alone, or as part of a number of syndromes (Czeizel, Dénes, & Szabó, 1973), and is the most common cause of pharyngeal stridor in infants. Retarded growth of the mandible results in micrognathia.

The stridor is produced by a mechanism similar to that responsible for snoring. *Due to the hypoplasia of the mandible, the tongue is displaced backward, obstructing or constricting the pharynx, and thus causing respiratory disorders.* The *pharyngeal stridor* is of the interrupted type because of changes occurring in the position of the backward displaced tongue. Within this type, the stridor may be characterized as stertorous, sawing, or snoring (Figure 7-5).

Micrognathia in lateral view can be ver-



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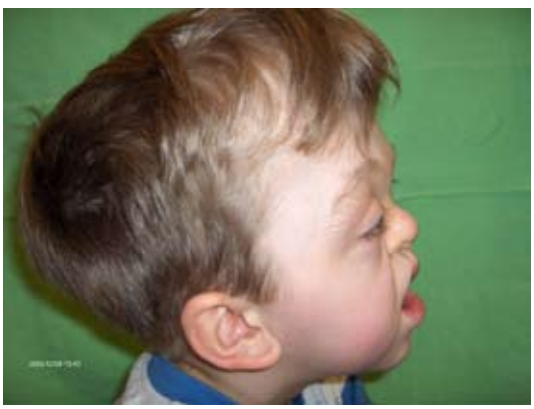

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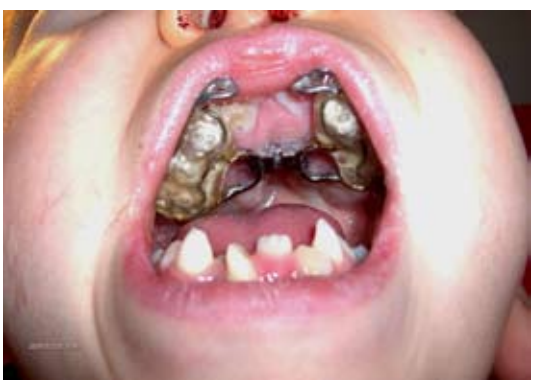

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FIGURE 7-4. A, B, and C. Apert syndrome in a 7-years-old child with bilateral craniofacial distractors, and before reoperation of bilateral choanal atresia.

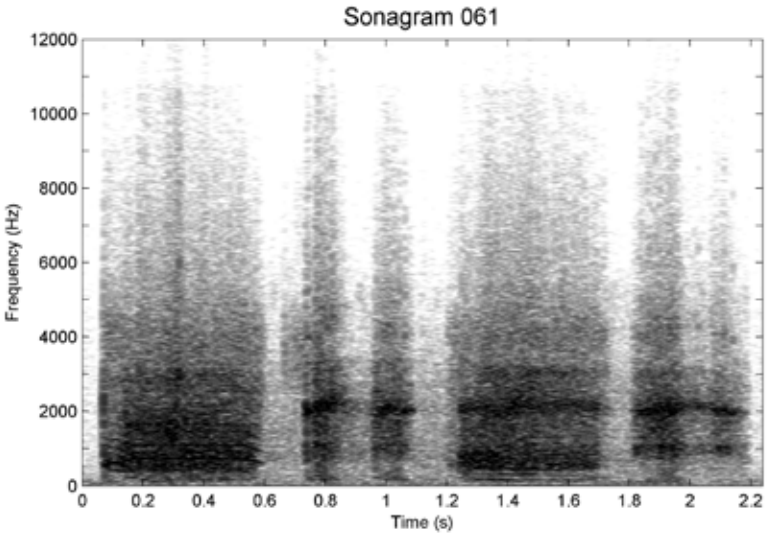


FIGURE 7-5. Micrognathia. Four-week-old infant. *History:* Common cold with difficult nasal breathing for 2 weeks. The choanae are passable. *Acoustic finding:* Typical pharyngeal stridor (inspiratory and expiratory). *Sonagram No. 61:* Harmonic structure strongly masked by noise; overtones between 450 to 1000, 2000 to 2600, and 3000 to 3500 Hz. On expiration there is a uniform masking by noise up to about 5500 Hz, where the spectrum is interrupted. The inspiratory phase consists of two parts and is much shorter than the expiratory phase. The auditory impression is of a high-pitched stridor because of the few prominent high-frequency overtones.

ified easily in general (Figures 7-6A and B), although the mandible of even normal infants is relatively smaller than that of adults. This fact as well as the borderline cases may make the diagnosis of micrognathia questionable lending special importance to the observation of pathologic sounds. The stridor in micrognathia is always noiselike, has no harmonic character, and only an occasional quasiperiodic structure can be observed. It usually decreases or disappears on subluxation of the mandible or if the tongue is elevated with the laryngoscope.

Observation (possibly polysomnography) and the *suitable position of the infant* (lying prone or sideways) *is enough action for most patients*, at minimum nasopharyngeal tube may be applied, but *in very severe cases* intubation or tracheotomy is recommended, similarly as in cases with Robin sequence (see next section), which may be life saving. Surgical maneuvers to advance the tongue, for example, glossopany, or mandible in the infant remain controversial (Isaacson, 2000). According to Mandell, Yellon, Bradley, Izadi, and Gordon (2004b) the use of *mandibular distraction osteogenesis can help* to avoid tracheotomy. The anatomic disproportion becomes settled generally spontaneously during the years; oral surgery correction therefore rarely comes up.

Micrognathia occurs not only as part of certain syndromes, especially Robin sequence, but may also be associated with other congenital malformations, for example, laryngomalacia. In the latter case the stridor may be either pharyngeal or laryngeal, or the two types may appear alternately (see Figure 8-1).

Robin Sequence

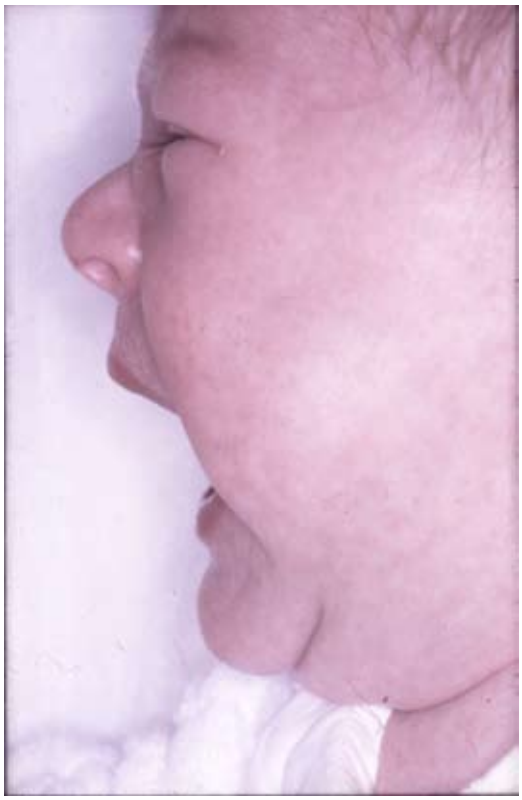
The incidence of this malformation is 0.08 to 0.09% in newborns (Czeizel & Hirschberg, 1997; Czeizel, Hirschberg, & Nagy, 1986; Czeizel, Hirschberg, & Tary, 1986). *The major features of this syndrome include micrognathia, a U-shaped palatal cleft (Figure 7-7), hypoplasia, and a backward displacement of the tongue.*

The classical triad was first described by Robin (1923). The natural proportions of the tongue, oral cavity, and mandible are altered, *the tongue has slipped backward causing difficulties in feeding, asphyxia, and pharyngeal stridor* (Hirschberg, 1973a). Robin sequence may occur as an isolated problem or as part of several congenital anomaly complexes and syndromes (Isaacson, 2000).

KEY POINT

The classical triad of Robin sequence is: micrognatia, U-shaped cleft of the palate, and backward placed tongue. The anomaly causes typical pharyngeal stridor in newborns and in severe cases respiratory distress. Prone position is mostly enough, other intervention (nasopharyngeal tube, intubation, tongue to lip adhesion, or later distraction osteogenesis) are rarely necessary. The closure of the palate is generally performed at the age of 10 to 12 months; preoperative polysomnography is recommended.

Stridor is produced in the same way as in micrognathia. It is usually high-pitched, *sawing, or snoring,* and can be observed in both inspiration and expiration (Figure 7-8). The more considerable the obstruction, the louder the stridor may become: but in general one can not draw any correct conclusion from the intensity of the



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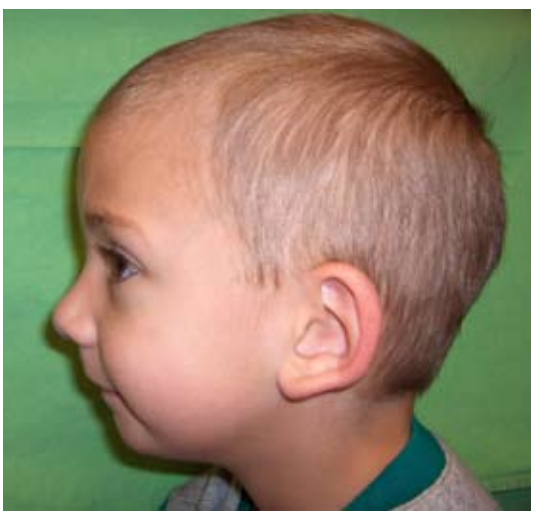

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FIGURE 7-6. Micrognathia in a 13-month (A) and a three-and-a-half-year-old (B) child.

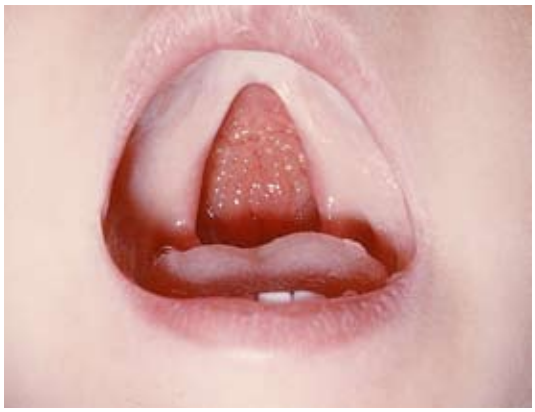


FIGURE 7-7. Typical U-shaped cleft of the palate in a child with Robin sequence.

respiratory noise alone as to the severity of the anomaly and the general condition of the infant. The cry is always clear and regular.

In typical cases the diagnosis can be established on inspection, whereas in formes frustes (where there is no cleft palate, merely a gothic palate) the acoustic finding may play a much more significant role. In newborns the first and most important task is ensuring unimpeded respiration. Prone posturing is mostly effective in the treatment of mild airway obstruction, according to literary data (Kirschner et al., 2003; Leung & Cho, 1999; Li et al., 2002) in 70 to 80% of the cases. When placement in a prone position fails to alleviate respiratory problems, some clinicians advocate the use of nasopharyngeal tubes. This is done by inserting the tube into one of the nostrils and fixing its other end behind the tongue base at the top of the larynx. This method not only ensures sufficient space for the passage of air within the tube but also prevents the tongue from falling backward (Benjamin & Walker, 1991). Only few infants require endotracheal intubation, due to severe respiratory distress, or a tongue to lip adhesion operation. Tracheotomy is indicated in about 3 to 5% of patients, but greater numbers are also given in the literature. Kirschner et al. (2003) reported that among 107 patients 4 required tracheostomy, and that tongue-lip adhesion successfully relieves airway obstruction that is unresponsive to positioning alone. According to Li et al. (2002), 6 patients needed tracheotomy among 110 children with Robin sequence. All these patients will later be able to be decannulated. With regard to feeding difficulties, patients may need nasogastric tube feeding, gastrostomy is not necessary. In a series of 117 cases of Robin sequence, Holder-Espinasse et al. (2001) found 48% isolated, 35% syndromic patients, and 17% with associated anomalies. The latter group had a poor long-term prognosis. For the management and prognosis of Robin sequence, the morbidity and mortality had been widely decreased through the teamwork of pediatrician, anesthesiologist, otolaryngologist, dentist, and plastic surgeon. In children with severe underdevelopment of the lower jaw, a new technique called mandibular bone expansion is now available. This technique also called distraction osteogenesis involves placement of an expansion device that is turned daily to slowly lengthen the jaw (Cohen, Simms, & Burstein, 1998; Mandell et al., 2004b; Tibesar, Price, & Moore, 2006). To avoid tracheostomy, Denny and Amm (2005) also recommend the progressive elongation of the mandible (distraction osteogenesis), to correct tongue ptosis, increase the passage of the pharyngeal airway, and correct micrognathia. A 5-

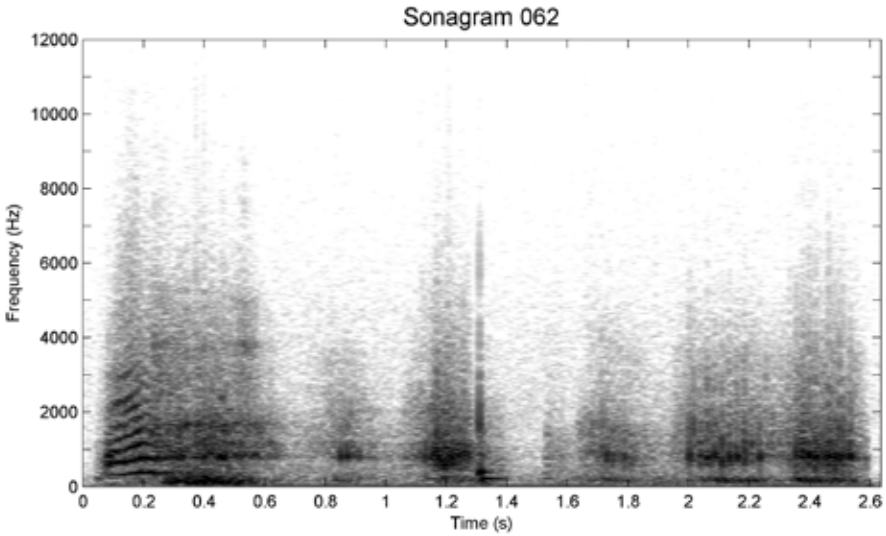


FIGURE 7-8. Robin sequence. Infant aged 4 months. *History:* Snoring, difficulty of inhalation and feeding from birth; cleft palate. *Acoustic finding:* Snoring pharyngeal stridor. *Sonagram No. 62:* The short introductory cry is followed by an interrupted pharyngeal stridor. The recurrent snoring phases are characterized by prominent, deep, confluent noise blocks. The quasiperiodic repetition of snoring cannot be seen because of the low-frequency values.

year clinical follow-up showed the operated mandible to maintain a normal shape and produce an undisturbed tooth eruption sequence. The authors conclude that *distraction osteogenesis is an effective alternative to tracheostomy in carefully selected neonates*. According to the publication of Smith and Senders (2006) one-third of 60 patients with Robin sequence were temporarily stabilized with a nasopharyngeal airway or endotracheal intubation. The remaining two-thirds of children, who failed positional therapy, required a surgical airway procedure. Four patients underwent mandibular distraction osteogenesis, resulting in successful decannulation or avoidance of tracheostomy. Thirteen patients underwent tracheostomy; the mean duration of tracheostomy-dependence was 17.0 months in isolated Robin sequence cases and 31.7 months in children with other anomalies. Successful decannulation by the age of 3 years was confirmed in 85% of patients.

Regarding the palatoplasty the general accepted rules for this surgery should be followed (Hirschberg & Gross, 2006). Before the closure of the cleft palate, we suggested polysomnographic examination (Hirschberg, 2004): if sleep apnea exists, or the oxygen saturation is not adequate, the palatoplasty should be postponed until these parameters are improved.

Cleft Palate (CP), Velopharyngeal Insufficiency (VPI)

According to a wide range of statistical data, the prevalence of the cleft lip/palate in different parts of the world is 0.5 to 2.2‰, whereas the occurrence of VPI cannot be exactly determined (Hirschberg & Gross, 2006).

The most common disorders of speech and voice in children with CP or VPI are

the following: retarded speech development, hypernasality, nasal escape, articulation disorders (mostly secunder), facial grimacing, and hyperfunctional phonation (dysphonia). The most important among them is hypernasality, the main acoustic properties of which was summarized as follows (Hirschberg, 1986; Trenschel, 1977): a relatively intensive fundamental frequency, decrease of the intensity of the first formant, dominance of noise components in the speech signal in general, the presence of (some nondefinite traces of) nasality around 500 (sometimes 1000) Hz in the spectrum, and lengthening of consonant duration.

KEY POINT

In infants with cleft palate (with or without cleft lip), the phonation is generally regular, and clear. We can demonstrate only traces of hypernasality in the cry with new diagnostic methods (nasometry, digital signal processing procedure). This result holds out hope to discover cases with latent velopharyngeal insufficiency in early infancy. The cleft should be operated on according to general accepted rules (see Hirschberg & Gross, 2006).

It has always been an open question whether increasing of the resonance, that is, hypernasality (hyperrhinophony) could be demonstrated already in the newborn's cry or first in conscious speech.

Bosma et al. (1965c), Massengill (1969), and Agnello, Hess, Mylin, and Hagerty (1973) could not find any alteration with spectrography in the cry of infants with CP.

Michelsson et al. (1975) have analyzed 52 phonations of 13 cleft palate neonates by sound spectrographic methods; 17 phonetical attributes were included in the study. The first signal after the pain stimulus was analyzed and compared with the crying of 75 normal babies of the same age. No change in the fundamental frequency, melody type, and duration of the cries was seen in association with these anatomic defects. Two of the characteristics studied, vibrato and the "tonal pit," occurred significantly more often in cries of the cleft palate infants than in cries of the control group.

Wermke et al. (2002a) studied the cry parameters of five patients with complete unilateral cleft lip, alveolar ridge, and hard and soft palate (UCLP) from birth to 9 months of age. The preliminary results of this study showed that the fundamental frequency (F_0) and the pitch perturbation quotient (PPQ) of spontaneous cries are influenced by UCLP and—according to the authors—a cry analysis might become a noninvasive tool for early detection or prediction of at-risk status for later speech and language acquisition in infants with cleft lip and palate.

The same was sought by the Budapest team: to detect as early as possible voice disorders (hypernasality) in infants with CP (Figure 7-9) and especially with latent velopharyngeal insufficiency (Figure 7-10): submucous cleft palate (Figure 7-11), occult submucous cleft palate, shortening of the velum, and anatomic disproportion. According to the data of Online Mendelian Inheritance in Man (OMIM, 2002), today more than 200 congenital anomalies are known, a part of which is some palatal defect. One of the most frequent cases among them is the velo-cardio-facial (VCF) syndrome (Sedláčková, 1967c; Shprintzen et al., 1978). As its name indi-

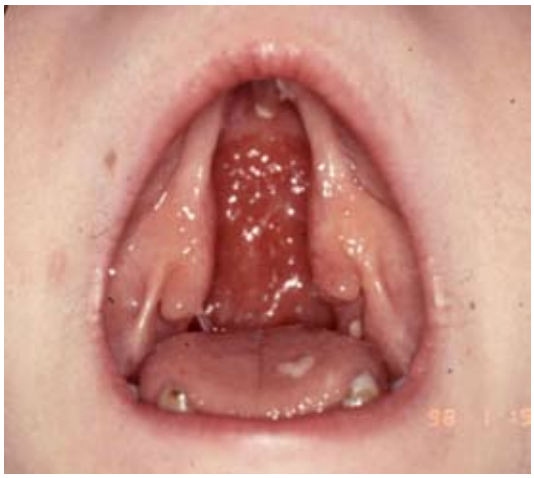


FIGURE 7-9. Large cleft of the palate (Case study, see text).

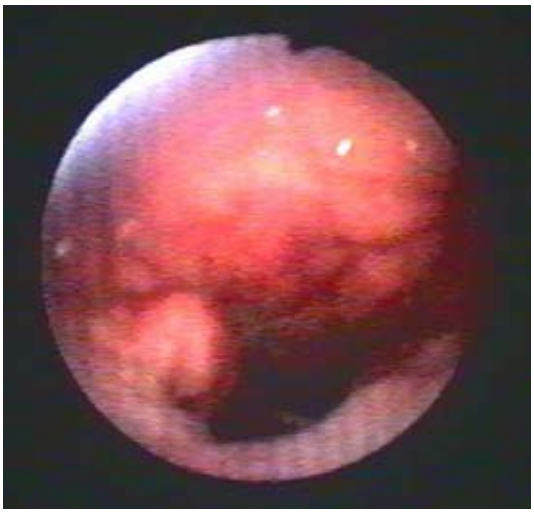


FIGURE 7-10. Nasopharyngoscopic picture about a velopharyngeal insufficiency. See the gap between the velum and the posterior pharyngeal wall during phonation.

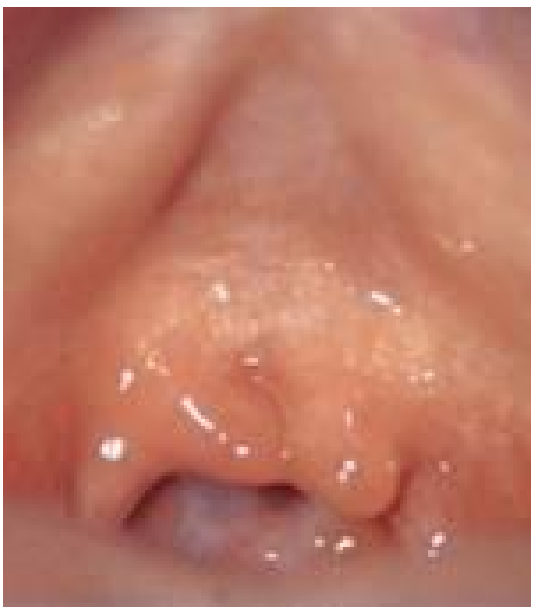
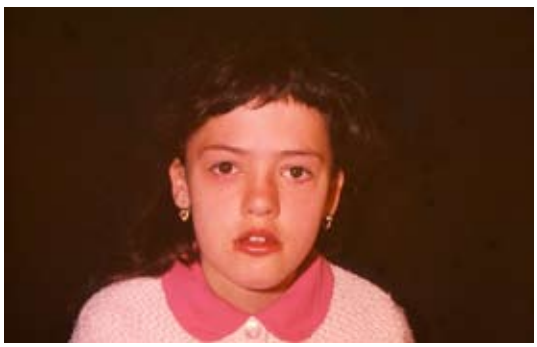


FIGURE 7-11. Submucous cleft palate. Child aged 17 months.

cates, the facial symptoms (flattened nasal root, upward lifted upper lip and philtrum, some deformity of the auricle) belong to this anomaly besides the cardiac and velar disorders. Chegar, Tatum III, Marrinan, and Shprintzen (2006) investigated 121 subjects with VCF syndrome: these patients showed significantly more asymmetry in the upper airways compared to the normal group (69% versus 20%, P = 0.01) with greatest differences seen with palatal motion, posterior pharyngeal wall size, and epiglottis shape. The physiognomy of the children suffering in VCF syndrome is typical (Figures 7-12A, B, C, and D), it is noticed at a later age and the hypernasality due to VPI is often misdiagnosed as hyponasality with faulty therapeutic consequences: repeated adenoidectomy may be performed (Hirschberg & Gross, 2006). For the sake of adequate management an early and correct diagnosis is essential. We wanted to gain evidence on whether the acoustic cues characteristic of cleft palate speech of children (the hyperrhinophony) can be revealed at an early age, that is, in infancy, on the basis of relevant properties of the cry. And if so, whether the surgery (palatoplasty) results in a change of the acoustic spectrum at that age.



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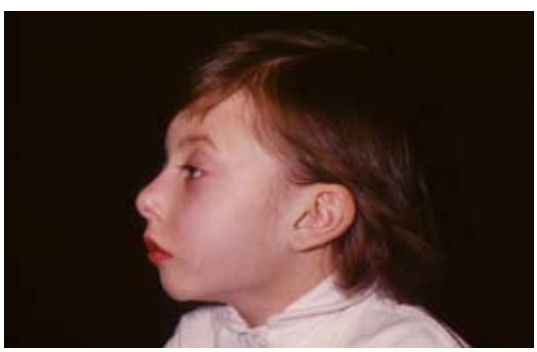

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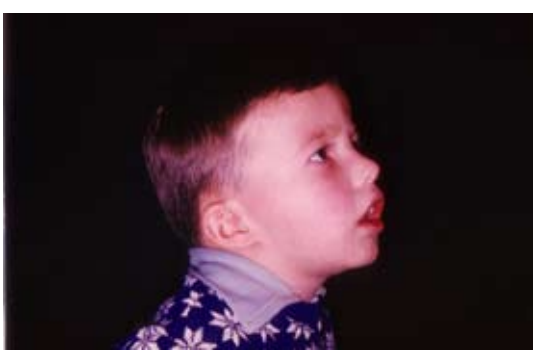


D

Fifteen infants between the ages of 7 to 14 months were examined prior to and five days after surgery by spectral analysis (Simon-Nagy, Szende, & Hirschberg, 1992). Results: the cry of cleft palate infants is usually clear (Figures 7-13A and B) and does not exhibit characteristic distinction of hypernasality in voice production. On the other hand, some data concerning the nasality component in cleft palate cry still may be heard. Differences between the voice production after operation and the cleft palate counterpart of the same infant are as follows: (1) The higher frequency constituents in the domain over 2800 to 3000 Hz of the cleft palate cry segment take a lower intensity value relative to the ones in the cry recorded after surgery; (2) the width and the relative intensity of the component about 1150 to 1300 Hz in the cleft palate version exceed the respective values of the voice produced by the child in the postoperative state (see Figures 7-13A and B).

To get more and reliable data about the characteristics of the cry in infants with CP, we introduced the application of nasometry in our study (Hirschberg et al., 2006). The preliminary results showed that the nasalance score, the grade of nasality of the cries in some cleft palate infants, is higher than in healthy infants. We have found ex-

FIGURE 7-12. Children with velo-cardio-facial-syndrome and consecutive hypernasality. Typical facial expression, faulty reminding the doctors of children with enlarged adenoid. The incorrect diagnosis of hyponasality due to adenoid vegetation resulted in repeated adenoidectomy in each of three children shown. The right diagnosis could albeit easily be established with the mirror test: the cold mirror setting under the nose become vaporous during phonation of vowels in cases of hypernasaliy due to VPI, but will not be vapory in cases of hyponasality due to nasal obstruction.

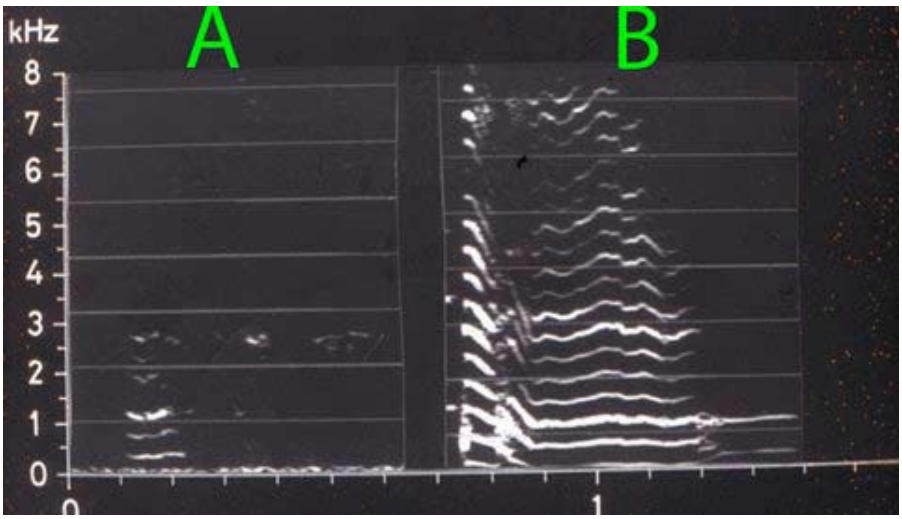
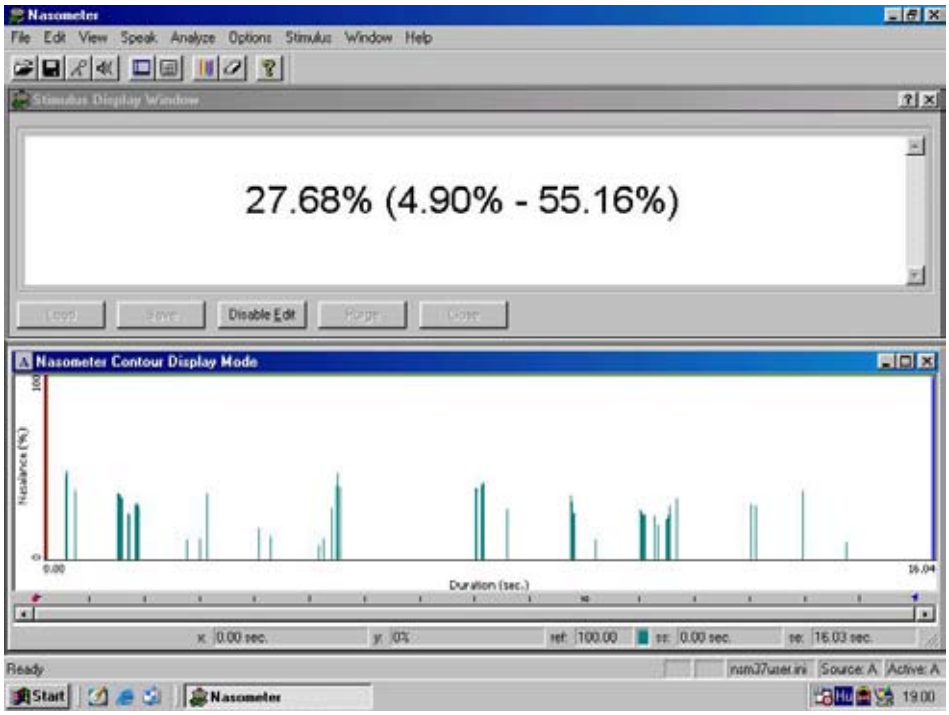


FIGURE 7-13. Comparison of sonagrams analyzed of the cries of an infants with cleft palate, aged 8 months, before (**A**), and after (**B**) palatoplasty.

tremely high degree of nasality values in a cleft palate baby, as well (Figures 7-14A and B); however, the differences between the cry of healthy and CP infants in the whole group studied were not statistically significant. Undoubtedly, the fact, that the adapted and previously used tests referred only to the speech of older children, making the investigation difficult because a necessary constant intensity level of the cry sound (about 60-70 dB) could not be expected in small infants. Besides, the condition of the subjects (whether tired or sleepy or hungry) and some of the circumstances of examinations were not always the same. Further investigations on a larger population with standard conditions are needed to have reliable results. *Our aim is to detect supposed hypernasality due to (occult) submucous cleft palate or latent VPI with nasometry as early as possible.* Not evident cases easily can be overlooked by routine inspection especially in infancy with severe consequences later on, for example, repeated adenoidectomy erroneously performed.

Hearing Impairment, Hearing Loss

The most significant symptoms of hearing loss in children are the following: the infant does not react to noise effects, the babbling does start, but gradually becomes poor, and speech development is late or does not appear on time. The tardily starting speech of a child with perceptive hearing impairment are characterized by the following factors: the speech may be loud or soft, the pitch is mostly high, sometimes low, the articulation is disordered, especially sigmatism exists, the accent is far from normative, the resonance—due to the lack of auditory feedback—increases; and hypernasality is a typical sign (Hirschberg, 2001, 2003). *The essence of the therapy in perceptive type hearing impairment is hearing aid (above 30 dB hearing loss), and speech therapy which is as successful as early as it begins.* The pre-



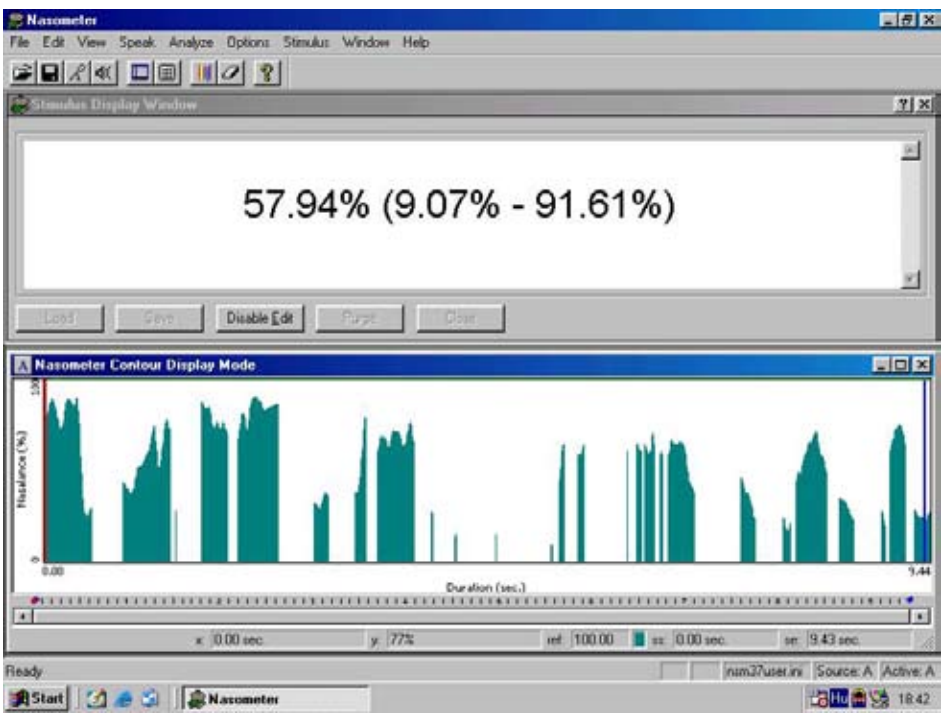


FIGURE 7-14. A. Nasogram of the cry of a one-week-old healthy infant. B. Nasogram of the cry of a one-week-old infant with cleft palate.

requisite of a good result is therefore the earliest hearing screening of the infant, the earliest correct diagnosis, as has been stressed recently in comprehensive books (Dillon, 2001; Roland, Marple, & Meyerhoff, 1997). If the diagnosis of a most severe hearing loss (deafness) is established and may be regarded as certain and definitive and an experienced surdologopedist could not aim at any result either, furthermore, no improvement is anticipated with the best hearing aid, cochlear implantation (CI) is indicated, before the age of 2 years. The first pediatric CI in Hungary (and also in former Eastern Europe countries) was performed by Hirschberg and coworkers (Farkas, Hirschberg, Simon-Nagy, & Katona, 1990; Simon-Nagy, Hirschberg, Katona, & Farkas, 1992) in a 3½-year-old preverbal deaf child in 1989 (Figure 7-15 shows the operative situation of the pediatric cochlear implant).

KEY POINT

In the speech of children with hearing impairment/loss hypernasality is one of the most typical symptoms. Our aim is to demonstrate the presence of increasing resonance as a consequence of the lack of auditory feedback in early infancy with noninvasive acoustic procedures. The objective audiologic methods (BERA, OAE) are not always available, but the infant cries everywhere and its voice sample may be sent on-line to an acoustic laboratory for evaluation.

The two pillars of diagnostic procedures in infants—apart from subjective observation—are the otoacoustic emission (OAE) and the BERA. These diagnostic instruments, however, are not always available and especially the OAE may give (occasionally) faulty results. Therefore, in recent decades, it seemed to be necessary to reveal the signs of the state of hearing impairment in the cry of the newborn infant with acoustic measurements.

Möller and Schönweiler (1999) proved with hearing experiments that it is possible for expert listeners to auditorily classify cries of hearing impaired and normal hearing infants, based on the characteristics of voice and melodic cry features. The cry of profoundly hearing-impaired infants are different regarding their perceived sound, rhythm, and melody. According to the authors the extracted signal parameters (obtained by means of new computer technique and neuronal networks) enable an automatic classification of cries by means of topologic feature maps which may be used later as a basis for an early supplementary diagnostic tool. Garcia and Garcia (2003) use also the neural network for recognition of the cry of normal and hard of hearing infants. The current results are promising and encouraging with an accuracy up to 97.43% in view of the reliability of evaluation. To this point it is worth mentioning, however, that Garbaruk (1998) has not found any difference in the vocalization of impaired and normal hearing infants aged 3 to 5 months when the author compared following acoustic characteristics of the two groups: duration parameters, formant frequencies, and fundamental frequency.

The Budapest team (Benyó et al., 2004; Farkas, Várallyay, Illényi, Benyó, & Katona, 2003; Várallyay et al., 2002a, 2002b, 2004a) examined the cry of normal and hard of hearing infants with digital signal processing (DSP) using a special method, which they developed called Smoothed

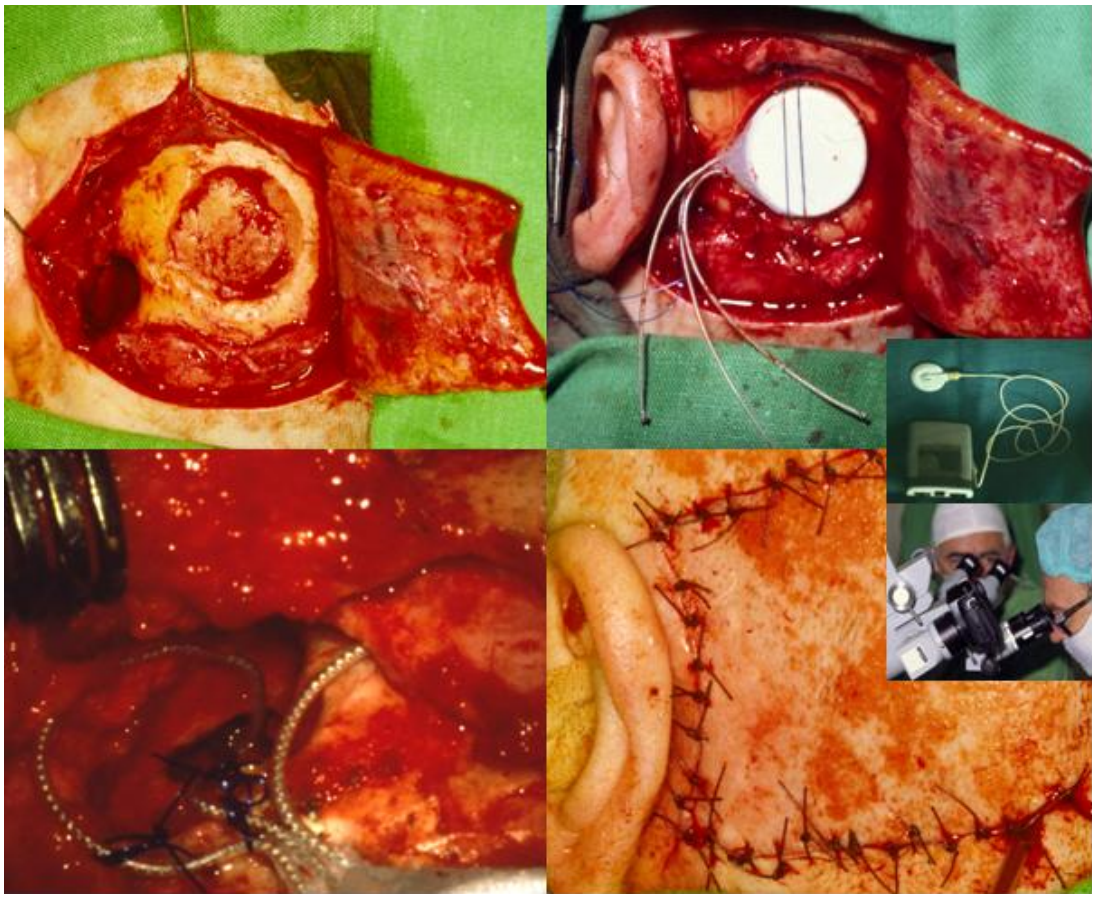


FIGURE 7-15. Historical pictures. Cochlear implantation of a three-and-a-half-year-old, preverbal deaf child in Budapest, 1989.

Spectrum Method (SSM) in 34 infants aged 1 to 15 months. The method is based on the spectral analysis but, simultaneously, combined with noise filtering and statistical processing. By comparing SSM with other methods such as cepstrum analysis or local maximum value detection the authors found preferences for this new method. They examined a total of 446 cry signals and 2,421 cry windows with a result of 97% efficiency. *Differences were found at the ratio of the dominant frequency and the fundamental frequency*. If the infant has normal hearing, this ratio is 3. If the infant has severe hearing impairment, the ratio of the dominant frequency and the F_0 is in most of the cases 2. The investigation is still ongoing with melody contour and the shape of the formants. The results enable to elaborate a rapid procedure for hearing screening of newborns with a noninvasive method.

Our other initiative is the application of nasometry to measure the resonance components of the cry in healthy and in hard of hearing infants (Hirschberg et al., 2006). *Preliminary results, however, cannot be regarded as reliable and definitive*. The na-

salance scores showed very diffuse values with respect of the nasalance score in healthy infants and in infants with hearing impairment, as well. It was, however, an interesting observation that the cry of hard of hearing babies demonstrated consequently different nasality scores with and without hearing aid. Further investigations are ongoing.

Cornelia de Lange (De Lange) Syndrome

This rare congenital anomaly, sometimes familial in occurrence (Beck, 1974), was first described by Cornelia de Lange. Its pathogenesis is still unclear (Leiber & Olbrich, 1972). *The syndrome is a genetic disorder present from birth.*

It is proper to make a remark here on the name of this syndrome. It has been generally accepted that the previously used term "Pierre Robin syndrome" should be labeled "Robin sequence" as Pierre is the first name of the first describer of the syndrome. Thus its use is unnecessary. Similarly, Cornelia de Lange's first name also may be omitted from the denomination of the anomaly and could be called: "De Lange syndrome."

The typical symptoms include a characteristic (clownlike) physiognomy, brachymicrocephaly, thick, fused eyebrows, hypertelorism, low front and back hairlines, a small, short nose, hypoplasia of the mandible, and deep-sitting ears. The hands and feet are small; syndactylia is common. There is also psychomotor retardation.

The stridor found in such infants is due to micrognathia. It is a pharyngeal stridor, which may be interrupted (Figure 7-16). In oligophrenia the stridor is even more marked. It diminishes if the infant is turned round to lie on its stomach. The cry of these patients is clear, but *the infant may have a low-pitched weak cry, too.*

Although there may be considerable variations in severity (Hesse & Kirchner, 1974), clinical diagnosis presents no problems, and the sound phenomenon is observed as a supplementary sign.

Treatment focuses on helping each child achieve his or her potential in terms of development and language, and medical care for physical problems. Infants benefit from early intervention programs for improving muscle tone, managing feeding problems, and developing fine motor ability (Tekin, 2002). Life expectancy is normal.

KEY POINT

De Lange syndrome is a genetic disorder. Its identification doesn't cause any problem on the basis its very characteristic outward appearance. The pharyngeal stridor due to micrognathia is an additional symptom. Management is really symptomatic.

Hurler's Syndrome (multiple dysostosis, gargoylism)

Hurler's syndrome is an inherited disease as an autosomal recessive trait, *and the most common form of mucopolysaccharidosis.* It was first described by Hurler (1919), therefore its name. Frequently it has a lethal outcome (Czeizel et al., 1973; Dieckhoff, 1974). Essentially, it is due to disorders in enchondral and periosteal ossification and to accumulation of pathologic substances (chondroitin, heparitin, and kerato-sulphate) in different tissues (spleen, liver, cornea, brain) caused by

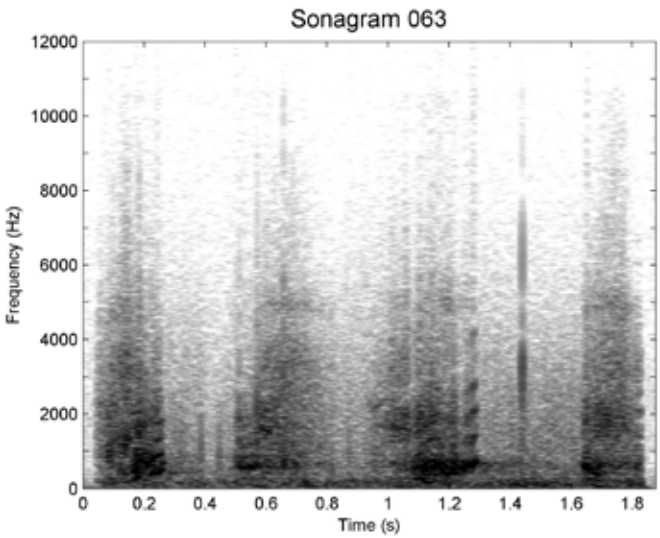
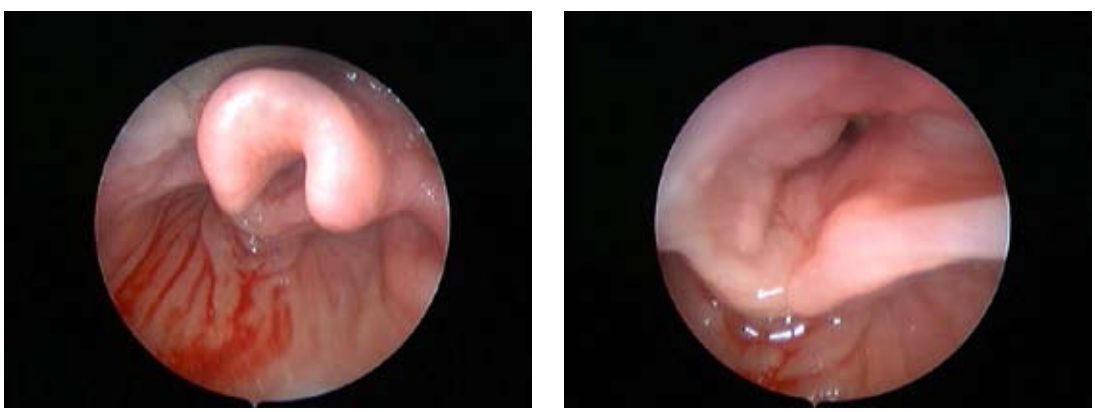


FIGURE 7-16. De Lange syndrome in an infant aged 10 months. *History:* Loud, noisy breathing from birth. *Acoustic finding:* Pharyngeal stridor, which is interrupted because of the movements of the backward displaced tongue. *Sonagram No. 63:* Confluent noise bands extending to 6000 Hz and interrupted at roughly the middle of the respiratory phases due to temporary occlusion of the pharyngeal cavity.

congenital enzymopathy. The disease also may affect the larynx, the consequences causing severe laryngeal chondral deformities (Figures 7-17A and B) with various sound phenomena.

The symptoms are highly variable. Newborn infants with this defect appear normal at birth but, by the end of the first year of life signs of impending problems begin to develop. *There is a progressive distortion of the whole body*, and a typical, grotesque mien, hence, the name gargoylism. There is also severe mental deficiency and deafness; the tongue is usually large, the lips are swollen, and the gingiva is hyperplastic.



A **B**

FIGURE 7-17. **A and B.** Larynx of an infant with Hurler's syndrome.

The disparity between the enlarged tongue and the oral cavity explains the frequently audible typical or interrupted pharyngeal stridor, resembling a gurgling sound (Figure 7-18). Increased salivation and oral congestion may also play a role in the production of this sound. In case of laryngeal chondral deformities hoarseness and laryngeal stridor also may occur.

Although the acoustic finding is variable according to the localization of the deformities, the appearance of the infant, the radiologic changes, and urinalysis leave little room for doubt as to the diagnosis.

The prognosis is bad. In cases of severe laryngeal anomalies tracheotomy must be taken into consideration. Death from heart disease generally occurs by the early teen years. Chemotherapeutics or recently enzyme laronidase are given to the child as a management possibility and bone marrow transplant may be considered as a successful procedure for potentially helping children with this disease.

KEY POINT

Hurler's syndrome is an inherited mucopolysaccharidosis with enchondral and periostal ossification. A progressive distortion occurs at the end of the first year with mental deficiency and hearing loss. Pharyngeal stridor may be observed due to enlarged tongue and in cases with laryngeal consequences hoarseness and laryngeal stridor also may be heard.

Mental Retardation, Mental Deficiency

Mental deficiency may accompany several congenital or acquired disorders. As it may be associated with a wide range of morphologic anomalies, the sound phenomena observed are highly variable. We refer here

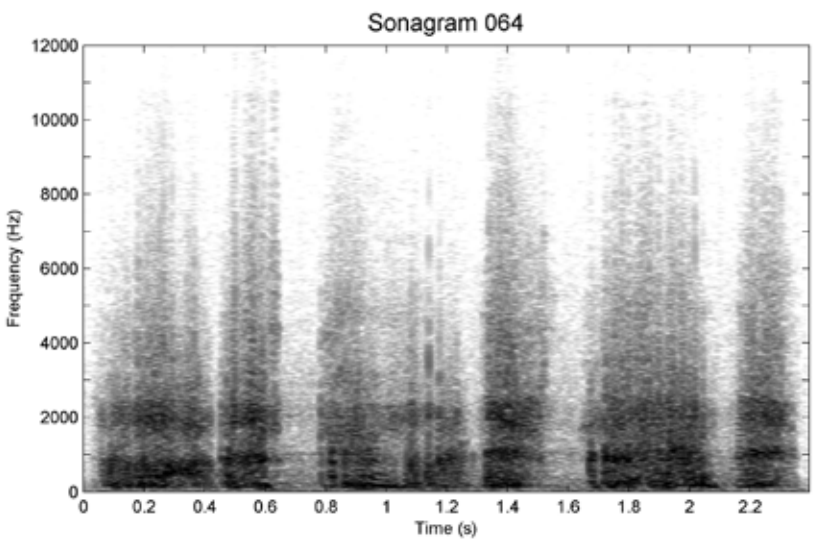


FIGURE 7-18. Hurler's syndrome. Infant aged 3 months. *History:* Upper respiratory catarrh and wheezy bronchitis since birth. *Acoustic finding:* Typical pharyngeal stridor having a gurgling-bubbling character. *Sonagram No. 64:* Expiratory, inspiratory, and expiratory stridor. Two distinct noise bands up to 1300 Hz and between 1600 and 3000 Hz. The noise bands are uninterrupted except for the inspiratory phase in the middle (0.95–1.1 s), which contains distinct vertical, columnlike noise blocks corresponding to the gurgling-bubbling part of the stridor.

only to the typical cry of the infant with Down syndrome (deep, creaking, pressed, interrupted, prolonged cry; see later section), to the variegated dysphonia in brain-damaged infants, and to the changes in vocalization occurring in hydrocephalus as a result of bilateral recurrent paresis (stridor-phonation, shrill or aphonic cry).

There are, however, some sound phenomena which may occur in several conditions independent of etiology. These may be regarded as signs common to all mentally retarded infants. As an example, *the interrupted snoring pharyngeal stridor* may be mentioned (Figure 7-19). It *is usually due to macroglossia and a backward displacement of the tongue*. Unusual articulation, or puffing, whirring, oropharyngeal, and labiobuccal sound production (Figure 7-20) also may be typical signs of mental retardation.

KEY POINT

Mental deficiency may have different types of etiology, the symptoms depend on the wide range of morphologic anomalies. Interrupted pharyngeal stridor due to macroglossia characterizes several conditions independent of etiology; it may be regarded as a common symptom in mentally retarded children.

Enlarged, Hypertrophied Tonsils, Adenoid Vegetation

The two usually occur jointly, although hypertrophy of the palatine and pharyngeal tonsils may also be found independently of each other.

Tonsillar hypertrophy (Figures 7-21A and B) was predominantly responsible for the stridor we observed and analyzed. *The sound is produced by air escaping through the narrow gap between the markedly enlarged, almost touching, tonsils. It is a typical noise, mostly undivided and audible in both respiratory phases. It is a characteristic lump-in-the-throat stridor* (Figure 7-22), sometimes also having rasping components. The pitch tends to be high. The duration of both inhalation and exhalation is prolonged. The respiratory noise is even more marked when there is an additional enlargement of the adenoids, due to the exclusively oral breathing.

The results of acoustical investigations performed by Slawinski and Dubanowicz-Kossowska (1993) indicate that as the nasopharyngeal tonsil increases, the dominant components of the respiratory sound spectrum shift toward a higher frequency range.

KEY POINT

Enlarged tonsils bring about a very typical, so called lump-in-the-throat stridor which is a noisy, undivided sound in both respiratory phases. The voice may be hyponasal, but if the strongly hypertrophized tonsils hinder the palatal movement, hypernasality also may occur. For the sake of a correct therapy, differential diagnosis between the two different resonance problems is essential. In children with OSAS (verified by polysomnography) early adenotonsillectomy is recommended.

In the cases of very large tonsils, which touch each other in the center line, sleep apnea may occur: in these children polysomnography is indispensable regarding the decision of a tonsillectomy at a very young age.

The big tonsils may make the voice hy-

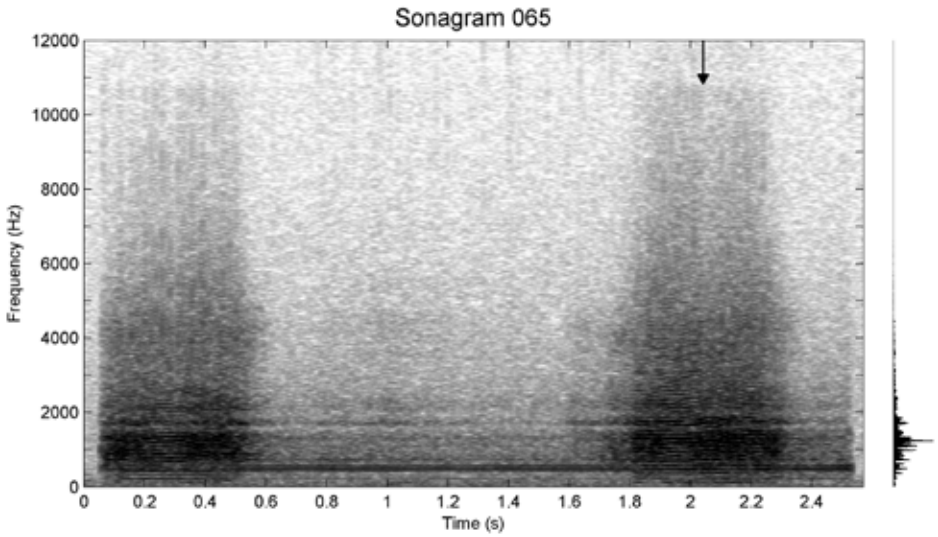


FIGURE 7-19. Mentally retarded infant aged 13 months. *History:* Prematurely born, repeated admissions to hospital, microcephaly, psychomotor retardation. Mouth can hardly be opened, the tongue slips back. Passable choanae. *Acoustic finding:* Snoring pharyngeal stridor due to a backward displacement of the tongue. *Sonagram No. 65:* Roughly divided noise up to 3000 Hz; a confluent noise block between 3500 and 6500 Hz. The snoring character is due to low-frequency components not visible in the sonagram.

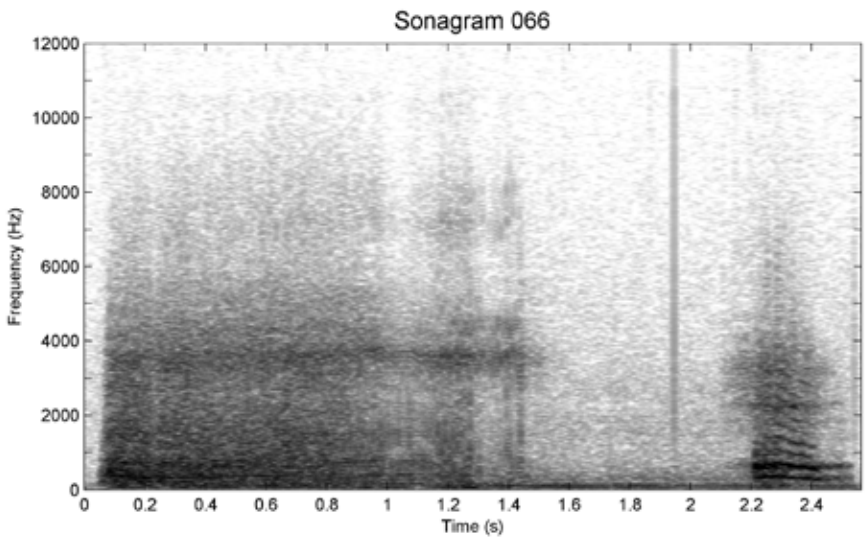
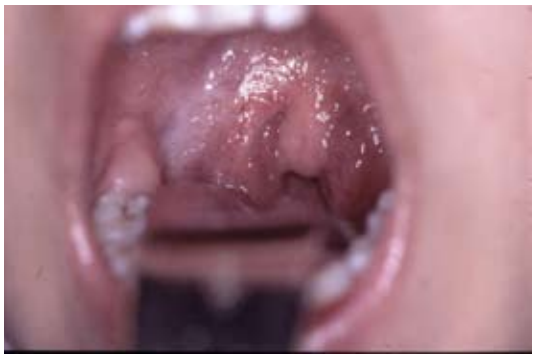
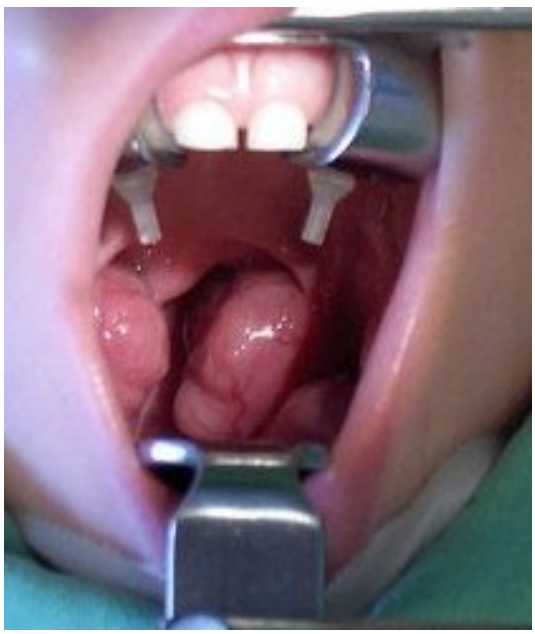


FIGURE 7-20. Two-year-old child seriously retarded due to environmental impacts. *History:* Coming from a wandering Gypsy family, admitted in an extremely neglected condition. Reported to have had "dyspnea" for 2 weeks. Microcephaly, severe motor and mental retardation; frightened and aggressive behavior. *Laryngoscopy:* No pathologic changes. *Acoustic finding:* Clear, inert phonation; labiobuccal, puffing-whirring articulation. *Sonagram No. 66:* A solid band of noise over 2500 Hz, the structure of which becomes simpler toward the end of the record. Intensive noise between 2500 and 6000 Hz, with a band of elevated intensity around 3500 Hz. The site of voluntary sound production is apparently the internal surface of the lips, and these sounds are associated with noises of higher frequency due to forceful blowing. This latter component does not appear when the child is short of breath. Between 2.0 and 2.2 s inspiratory phonation of regular acoustic structure.



A



B

FIGURE 7-21. Enlarged tonsils in a child aged 6 (A) and 7 (B) years, respectively.

ponasal; if, however, the very large tonsils hinder the movement of the velum, hypernasality also may occur. In these cases, regarding the surgery, close cooperation between the surgeon (otolaryngologist) and a phoniatrician or experienced speech pathologist is recommended. It may be all the more important as in cases with velocardio-facial syndrome (VCFS) the shortening and drawing up of the philtrum with the consequence of a protruding and so visible upper set of teeth (see Figures 7-12A, B, C, and D) may give the faulty impression of an open mouth (Hirschberg & Gross, 2006). The differential diagnosis may be difficult; thus, the proper evaluation of the voice is essential, occasionally combined with nasometry (Hirschberg et al., 2006).

Disorders Affecting the Larynx

Conditions discussed in this section include those affecting the supraglottic, the glottis, and the subglottíc areas. Nervous and myogenic disorders characterized by pathologic sounds which are produced in the larynx but cause no demonstrable local changes, are also discussed.

Epiglottitis (supraglottic inflammation)

In young children, this condition develops extremely rapidly, within an hour or two. Since the widespread introduction of the *Haemophilus influenzae* type B (Hib) conjugate vaccine about 13 years ago there has been a dramatic reduction in its incidence (McEwan, Giridharan, Clarke, & Shears, 2003). According to Faden (2006) the admission rate for acute epiglottitis declined tenfold in the past 27 years. In the immunized population streptococci are the most common cause of bacterial epiglottitis. Children are usually affected between the ages of 2 to 6.

Epiglottitis is an acute inflammation of the supraglottic part of the larynx, with a deep red, swollen, and dry mucosa. The epiglottis may be thickened, infiltrated and "clublike" (Figure 7-23). As the infection extends over the whole supraglottic lar-

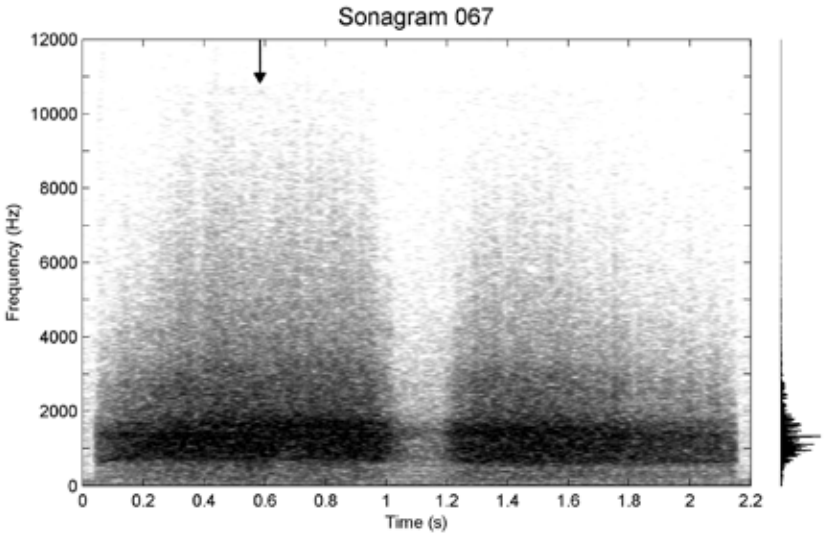


FIGURE 7-22. Hypertrophy of tonsils and adenoid vegetation in a two-and-a-half-year old child. *History:* Sleeps with mouth open and snores; mouth is held open during the day, too. *Acoustic finding:* Lump-in-the-throat stridor, loud inspiration and expiration. *Sonagram No. 67:* Inspiration between 0.1 and 0.9 s; expiration between 1.0 and 1.8 s. Between 600 and 2500 Hz dense, confluent, and intensive noise band associated with other, less marked components, for example, around 4500 Hz. Masking by noise at inspiration throughout the spectrum. The lump-in-the-throat stridor occasionally creates the impression of being high-pitched due to the high-frequency noise component. The duration of both respiratory phases—mainly that of inspiration—is prolonged.

ynx, would be more aptly labeled as supraglottitis (Sie, 2000). The vocal cords are intact and also the subglottic area remains uninvolved.

Dysphagia, a severe throat pain, and difficulty in breathing are the major symptoms in addition to fever. The characteristic outward features include a frightened look, facial expression reflecting pain, the tongue hanging out, and ample salivation. These children are usually quite anxious; they often are found sitting in the "sniffing" or "tripoid" position in an effort to maintain their airway (Sie, 2000).

Breathing is deep, slow (thus maintaining a better O_2 supply than with rapid breathing), loud, and noisy. *Respiratory noises are high-pitched and sharp. Stridor is audible both on inspiration and expiration*

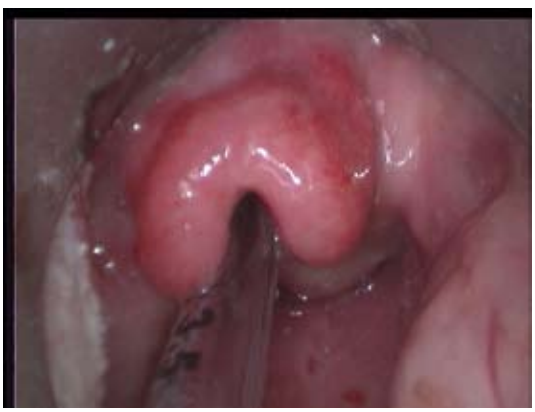


FIGURE 7-23. Acute epiglottitis.

(as described also by Beckmann, 1970), but is more marked in inhalation. The duration of inspiration increases. *Although this cannot be proved, we have the impression that breathing is painful: the inhaled air irritates the extremely sensitive, swollen*

supraglottic area (Figure 7-24).

The cry is clear because the process does not involve the vocal cords. Shah, Roberson, and Jones (2004), however, characterize the phonation as "hot potato voice." There is no barking cough because the subglottis is unaffected and this is important for the differentiation of this condition from stenosing laryngotracheitis.

KEY POINT

Epiglottitis is a rapid inflammation of the supraglottic area; the glottis and the subglottic area are unaffected. This situation explains the sound phenomena to be heard: a high-pitched and sharp, noisy, inspiratory and expiratory stridor, whereas the cry is clear and there is no cough. Since the introducing of vaccination the frequency of occurrences of epiglottitis decreased significantly.

Occasionally the diagnosis may be established by pressing down the root of the tongue carefully, when a swollen, inflamed epiglottis can be seen. Intraoral examination, however, should be performed only if equipment is available to secure the airway immediately (Sie, 2000) because an apnea may occur. Mostly lateral neck radiograph with lateral soft tissue neck film and expeditious direct laryngoscopy is performed. The stridor described may play a major role in eliciting suspicion of this diagnosis.

In the management intubation (for an average of 3.5 days, according to Shah et al., 2004), intravenous antibiotics and steroids are important. The most commonly administered antibiotics are ampicillin/sulbactam and ceftriaxone.

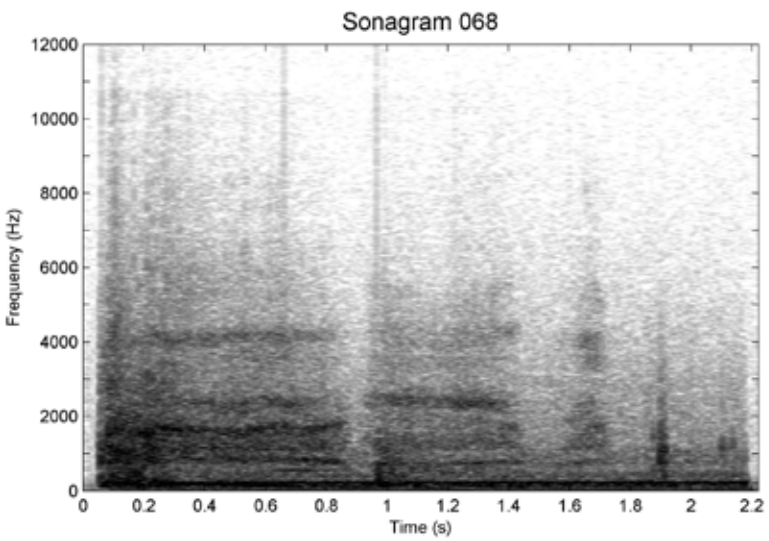


FIGURE 7-24. Acute epiglottitis in a child aged 3 years. (For the history and physical findings see legend to Figure 6-36). *Acoustic finding:* Loud, noisy, prolonged inspiration and expiration. *Sonagram No. 68:* Painful inspiration, then expiration. Prominent noise components between 500 and 1500 Hz; there are further, well-defined noise components at 2200 Hz, and during inspiration also around 4000 Hz. This stridor closely resembles the lump-in-the-throat stridor in its acoustic attributes except for the arrangement in bands of its dominant noise component. Painful and prolonged inspiration, and expiration with sharp, high-pitched respiratory noises. There is an attempt to swallow at the end of the record.

Acute Laryngitis

Here we list those changes in which the vocal cords are inflamed, swollen, and thickened due to grippe or acute upper respiratory infection, but there is no crust or fibrin formation, the subglottis is not involved, and there are no marked pathologic changes in the trachea either, apart from minor catarrh. *This is usually secondary to a viral upper respiratory tract infection, caused by parainfluenza virus* generally in fall and spring, but it may be caused by a bacterial organism, most likely Streptococcus pneumoniae or Haemophilus influenzae. Lately, gastroesophageal reflux (GER) has been implicated in the pathogenesis of the so- called reflux laryngitis, which is now a well-recognized phenomenon in children (Contencin et al., 1995; Silva, 2000). Gastroesophageal reflux is common in infants, but usually resolves spontaneously by 12 to 18 months (Saigusa, H., Niimi, Saigusa, U., & Yagi, 2001). Although the correlation between gastroesophageal reflux disease (GERD) and the extraesophageal manifestations has been well established, a cause-and-effect relationship has yet to be definitively elucidated (Gurski, da Rosa, do Valle, de Borba, & Valiati, 2006). The judgment of the correlation between GERD and laryngeal symptoms is controversial. According to Andrieu-Guitrancourt, Dehesdin, Le Luyer, Fouin, and Peron (1984) GER may be responsible for the onset of recurrent laryngitis in children. On the other side, Joniau, Bradshaw, Esterman, and Carney (2007) stress that at the moment there is no reliable means to confirm reflux of gastric juice in patients with suspected reflux laryngitis. Saigusa et al. (2001) reported 4 pediatric cases with unusual laryngeal disorders, especially posterior glottic lesion, induced by gastroesophageal reflux without other causal diasease. The diagnosis of laryngopharyngeal reflux is usually made on the basis of presenting symptoms and associated laryngeal signs including laryngeal edema and erythema (Farrokhi & Vaezi, 2007a). In our opinion, the etiologic factor of GERD may be accepted in cases (especially with posterior laryngitis, see Figure 7-25) when the patient could not be managed by routine laryngologic treatment, only by empiric therapy with proton-pump inhibitors (PPI). This latter is now considered the initial diagnostic step in patients suspected of having GERD-related symptoms (Farrokhi & Vaezi, 2007b). The PPI therapy, however, could not be overestimated (Jaspersen, 2006).

Since the primary involvement in cases of laryngitis is that of the vocal cords, the acoustic concomitant is disturbed primary sound production. The cry is veiled or hoarse (Figure 7-26). The veiled sound differs from the hoarse cry only quantitatively, depending on the extent to which the regular

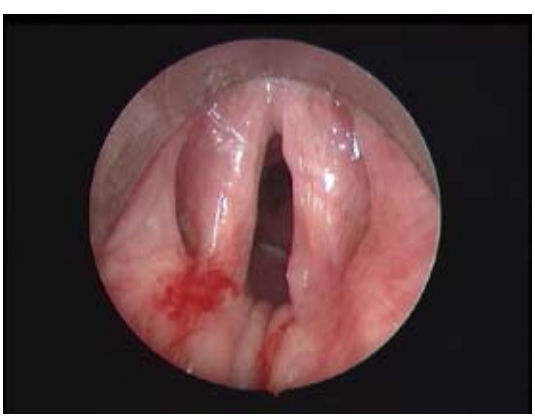


FIGURE 7-25. Gastroesophageal reflux with laryngitis.

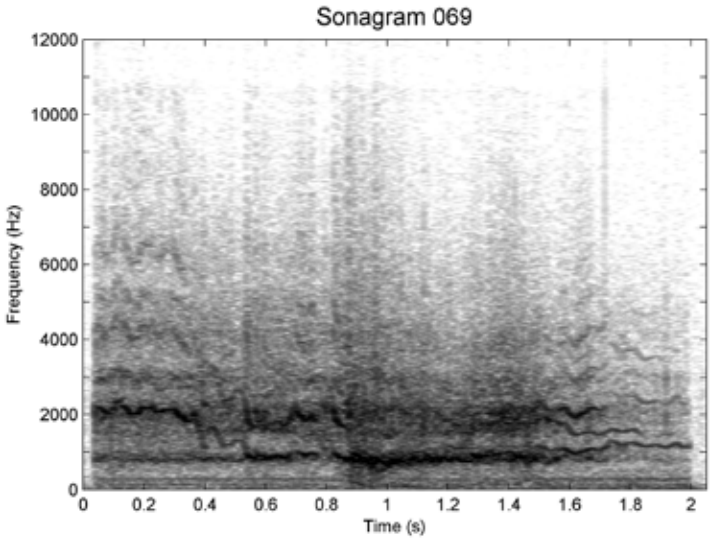


FIGURE 7-26. Acute laryngitis. Child aged 27 months. *History:* Hoarseness for a week. *Laryngoscopy:* Strongly inflamed, slightly edematous laryngeal mucosa. Mobile, inflamed vocal cords with uneven margins. Subglottis, trachea free. *Acoustic finding:* Hoarse cry. *Sonagram No. 69:* Due to the restricted mobility of the vocal cords the periodic vibrations required for a clear cry are absent and there are clearly divided noise bands in place of the harmonic structure at 700 Hz, between 2000 and 2500 Hz, at 3200 Hz, and between 4000 and 4500 Hz. The latter noise band is, however, rather vague. Hazy, continuous masking by noise. The only manifest attribute of the crying character is the duration of the sound phenomenon. Totally disintegrated formant structure. Acoustically, there are signs of grave hoarseness, which was also observed auditorily.

periodic vibrations of the vocal cords are impeded. As a result of the inflammatory edema, the vibrations of the mucosal surface are altered, and the regular acoustic structure is disturbed. The extent of this disturbance depends on the strength of the irregular vibrations. *There is no stridor*. Catarrhal coughing may occur, but the cough does not sound hollow or barking in the absence of subglottic swelling.

The patients are in relatively good general condition, and have only a low fever. The diagnosis can be established on grounds of the clinical symptoms and the cry, and confirmed by laryngoscopy.

Cold humidification should be ensured for all patients. Spontaneous resolution usually takes place but antimicrobial therapy may be indicated in cases that have a protracted course or accompanied with bacterial infection and purulent postnasal drainage which may be characteristic of sinusitis (Myer & Cotton, 1988). Voice rest is recommended for approximately 7 to 10 days, if possible, to older children.

KEY POINT

Laryngitis acuta is a viral infection involving the vocal cords mostly accompanied by low fever. The cry of the infant is veiled or hoarse; there is no stridor. Spontaneous remission usually takes place in 7 to 10 days. In prolonged cases, when the hoarseness has a longer duration, laryngoscopy is recommended.

Along with the healing of the process, sound production also improves.

The hoarse cry first becomes veiled, then clears up completely. The improvement is also reflected by the acoustic pattern.

Laryngeal Croup (acute fibrinous laryngitis)

The meaning of the term "croup" is vague in professional use as authors and also *physicians in everyday practice refer to different diseases by this expression*. According to some authors (Malhotra & Krilov, 2001) the term "croup syndrome" covers a group of diseases that vary in anatomic involvement and etiologic agents, and includes laryngotracheitis, spasmodic croup, bacterial tracheitis, and laryngotracheobronchitis. Others, like, for example, Brown (2002), take the view that croup is a common pediatric respiratory illness involving inflammation and narrowing of the subglottic region of the larynx, frequently precipitated by viral infections. Leung, Kellner, and Johnson (2004) consider viral croup and laryngotracheobronchitis to be synonyms both labeling one and the same clinical syndrome caused by various viral agents, and characterized by varying degrees of inspiratory stridor, barking cough, and hoarseness as a result of laryngeal and/or tracheal obstruction. In the chapter on Infectious and Inflammatory Disorders of the Larynx and Trachea in the textbook *Pediatric Otolaryngology* (Wetmore et al., 2000), Sie (2000) also includes viral croup under the title: "Laryngotracheal bronchitis."

As is well known, the upper respiratory viral infection does not respect the borders of every single anatomic region, at least in the majority of cases. The inflammation frequently spreads over the subglottis and over the trachebronchial tree from the glottis. Thus, it would be correct to treat also the laryngitis subglottica (pseudocroup) and laryngotracheitis in the section "laryngeal croup." However, in our book, we separate various distinct types of "croup syndrome" disease along the lines of the anatomic regions of their localization, that is, larynx, subglottis, tracheal, and bronchial affections, all the more relying on this way of classification because the several sound phenomena discussed could thus be interpreted better. This way of looking at things seems to be more advantageous also in teaching, even for the sake of recognizing correctly several distinct sound phenomena of the infant on each level of the airway.

In the case of laryngeal croup hoarseness or aphonia dominates. If the subglottis is involved and narrowed (laryngitis subglottica = pseudocroup), inspiratory stridor is the leading symptom: the cry is clear, at least veiled, and barking cough may occur; in tracheobronchial stenoses biphasic or expiratory stridor is typical, and hollow coughing (and possibly crying) sound may be observed.

Laryngeal croup, discussed in this chapter, used to be a common concomitant of diphtheria. Nowadays it is seen less frequently and occurs as a part of certain other infectious diseases such as influenza or morbilli. It occurs in young infants the most. Subglottic laryngitis and laryngotracheitis are discussed later.

In the case of laryngeal croup, the vocal cords are less red and less severely inflamed than in acute laryngitis and are *covered with a whitish, semolina-like, fibrinous exudate. This coat may extend beyond the margins of the vocal cords medially, preventing their perfect closure during phonation*. The

laryngeal change may be accompanied by tracheitis, but often the trachea is entirely free of inflammation.

As compared with catarrhal laryngitis, the quality of the voice is poorer in this condition. *Hoarseness is very marked, occasionally the cry breaks, and not infrequently there is aphonia* (Figure 7-27). The cause of these pathologic sound phenomena is the generation of noise by the floating fibrin particles extending beyond the vocal cord margins, or a very thick fibrinous coat strongly restricting the closure of the vocal cords in case of aphonia. Stridor, if it appears, is slight. Cough is not a characteristic symptom.

The diagnosis can be clarified by laryngoscopy. Removal of the exudate during direct laryngoscopy may result in a dramatic improvement of hoarseness. The clearing up of the voice can be readily demonstrated in the sonagram (Figures 7-28A and B).

KEY POINT

Laryngeal croup formerly was a consequence of diphtheria, at present it occurs due to influenza in young infants. The vocal cords are covered with fibrinous exudates: this coat may extend beyond the margins. These changes unambiguously explain the voice disorder: the cry is very hoarse, also aphonia may occur. Removal of the coats using direct laryngoscopy results in immediate improvement in the phonation and also the breathing will be easier.

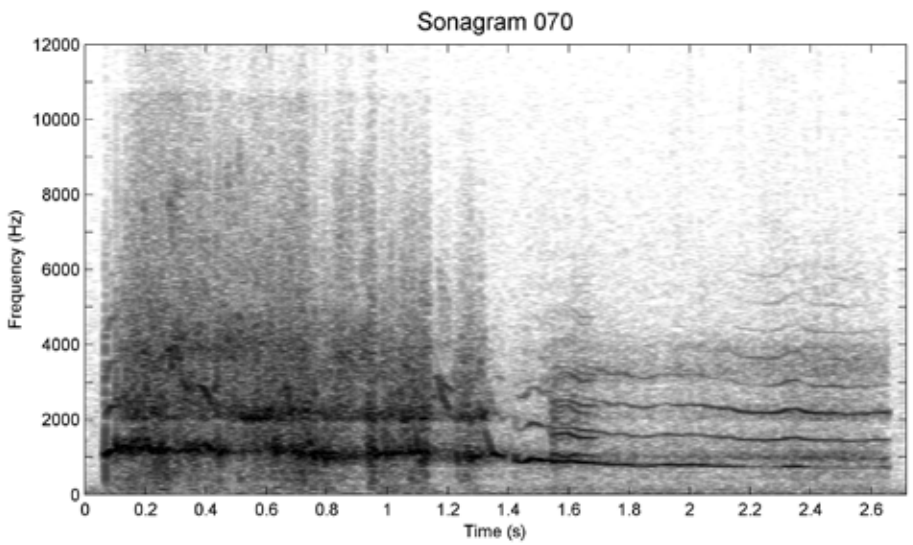


FIGURE 7-27. Laryngeal croup. Infant aged 7 months. *History:* Following symptoms of grippe, increasing hoarseness and aphonia of a few days' standing. *Laryngoscopy:* Whitish, fibrinous apposition on the vocal cords, moderate inflammation of the laryngeal mucosa. The subglottis is free. *Acoustic finding:* Excessive hoarseness. Occasionally the voice clears up or there is a break in the cry; total aphonia eventually. *Sonagram No. 70:* Regular, but scarce harmonic structure with the overtones as far as 1000 Hz apart. Over very brief segments (1.4–1.6 s) additional overtones appear. The spectrum extends to 4000 Hz. Marked masking by noise, with energy maxima at approx. 1300, 2400, 4200, and 4800 Hz; there is a dominant noise block between 2300 and 10000 Hz, in the time range of 0.02 to 1.15 s. Due to the fibrinous exudate, the incomplete harmonic structure disappears for shorter or longer periods giving way to aphonic phases which are slightly tonelike owing to a fairly regular arrangement of the noise.

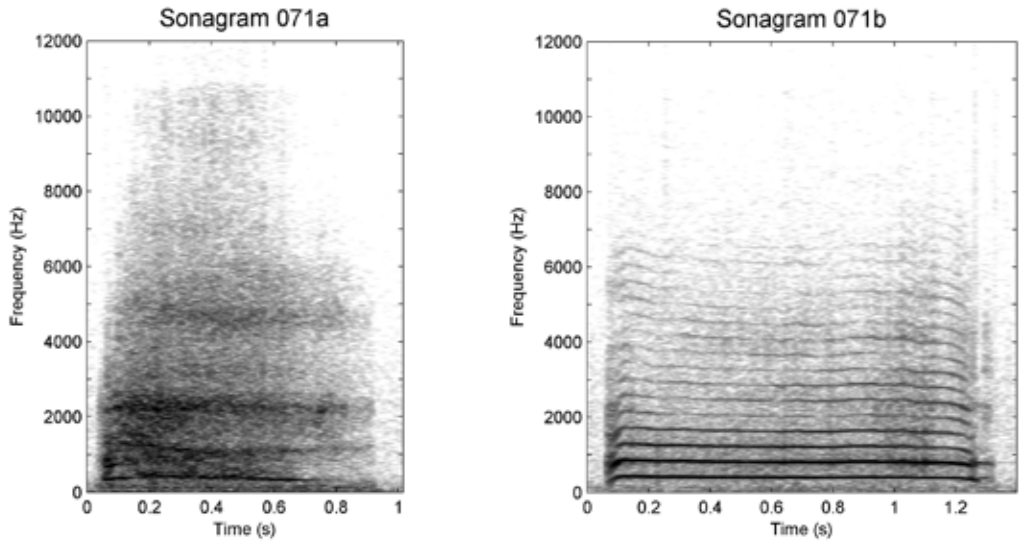


FIGURE 7-28. Laryngeal croup. Infant aged 7 months. *History:* Hoarseness and subfebrility for a day. *Laryngoscopy:* Moderate, diffuse inflammation of the laryngeal mucosa. Mobile, noninflamed vocal cords with gritlike, whitish, fibrinous coating on both sides, extending over the margins of the vocal cords, which thus cannot close during phonation. Subglottis, trachea normal. *Acoustic finding:* Hoarse cry (A) which clears up immediately after the direct endoscopic removal of fibrin (B). **A**. *Sonagram No. 71a: A.* No harmonic structure recognizable, amorphous noise throughout the entire width of the spectrum, with more intensive components around 2500, 4500, 6000 Hz, and further up to 10000 Hz. Hoarse sound. **B**. *Sonagram No. 71b:* Regular harmonic structure, with the overtones about 400 Hz apart. Vague masking by noise, mostly above 3500 Hz. Slightly veiled, regular cry.

The medical therapy depends on the bacterial finding. Besides antibiotics, cold humidification, oxygen, nebulized budesonid, racemic epinephrine, and occasionally steroids are recommended (Klassen et al., 1998). Intubation is necessary only for severe respiratory distress and it may be applied only for a short period.

Laryngeal Thrush, Laryngeal Mycosis, Fungal Laryngitis

The laryngoscopic appearance of this alteration is essentially the same as that seen in acute fibrinous laryngitis. Partly confluent, whitish thrush colonies, surrounded by a halo of slight superficial inflammation (edema, hyperemia) may be seen on the vocal cords or sometimes granulationlike growths on other part of the larynx (Figure 7-29).

The disorder of vocalization is caused by the membranelike fungus colonies which make the vocal cord margins uneven and protrude into the glottic lumen. The cry is hoarse, and aphonia, too, may occur in some cases. If the granulation tissue is on another part of the larynx, the sound phenomena are not so characteristic.

KEY POINT

In cases of laryngeal thrush the laryngoscopic picture is similar to that of laryngeal croup: whitish thrush colonies cover the vocal cords. In these cases the cry is hoarse. Mycologic examination confirms the diagnosis.

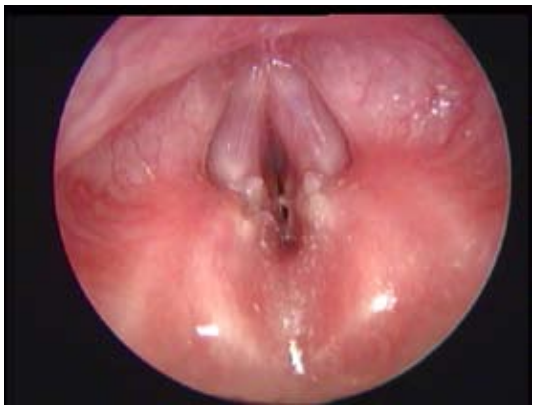


FIGURE 7-29. Candida of the larynx.

In the case of hoarseness this condition should be considered if thrush colonies are visible in the oral cavity. Superficial fungal infection of the mucous membrane may, however, occur isolated to the larynx with no oral or oropharyngeal manifestations, but first of all in adults (Sulica, 2005). Burton, Seid, Kearns, and Pransky (1992) presented a case of an otherwise healthy infant with severe croup who was hospitalized and treated with both steroids and antibiotics. A relapse in her symptoms led to the diagnosis of *Candida* laryngotracheitis. Therefore, the authors recommend close monitoring of patients with croup treated aggressively with steroids and antibiotics: steroid should be limited to 24 hours with antibiotics reserved for patients with signs of bacterial infection. Wang, Liu, Huang, Huang, and Wu (1997) reported a case of laryngeal candidiasis in an immunocompetent infant: the diagnosis was obtained by direct fibreoptic laryngoscopy with specimens submitted for culture. Immune-compromised patients are prone to fungal infection: when the body's immune system is suppressed, certain fungi that usually do not cause infection may penetrate the natural blood and tissue barriers and cause infection of the larynx (The Voice Problem Web Site, 2004).

The diagnosis can be established by laryngoscopy and mycologic examination. Following removal of the thrush colonies by direct laryngoscopy the voice clears up, which can be readily documented by spectrography (Figures 7-30A and B).

Antimycotic therapy may be indicated.

Changes Following Prolonged Intubation, Acquired Laryngeal Stenosis

We have to deal with prolonged intubation if the tube is left in place in the airways for more than 24 hours at a time. Statistics give the incidence of local complications as approximately 4% (Tonkin & Harrison, 1971). The duration of the prolonged intubation is, however, defined very variably in the literature. With improvement in medical care, many more premature intubated infants are surviving, increasing the number of infants with acquired subglottic stenosis (Zalzal, 1989). It has been reported in almost 7% of all intubated infants seen in the intensive care unit, and this figure may underestimate the real incidence (Myer & Cotton, 1988). Kim and Baek (2006), on the basis of analyzing 249 cases with laryngotracheal stenosis, stress with good reason that it is very desirable that the duration of endotracheal intubation be limited to less than 20 days.

Of the early complications (those following extubation immediately or shortly afterward) lesions of the vocal cords like subepithelial hematoma, glottic edema (Figure 7-31), exulceration, granulations (Figure 7-32), and pseudomembrane formation are

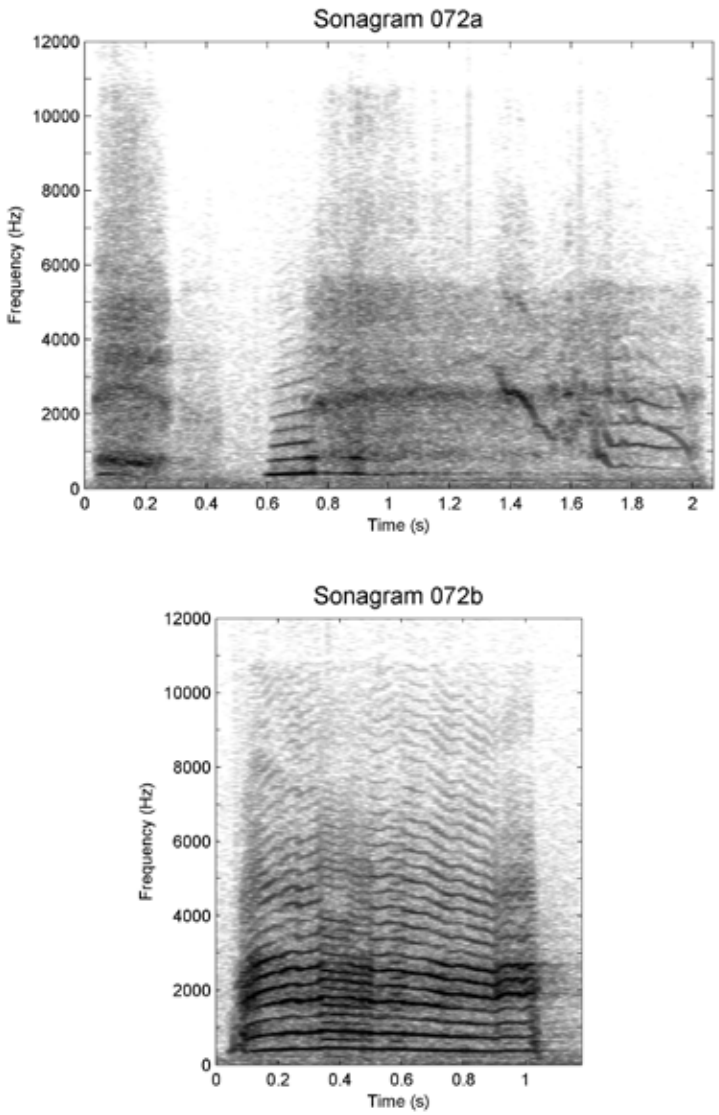


FIGURE 7-30. Laryngeal thrush (verified by mycologic examination). Infant aged 9 months. *History:* Gradual development of hoarseness over 2 days. *Laryngoscopy:* Moderately inflamed laryngeal mucosa, mobile vocal cords, with whitish spots on the middle and anterior thirds and on the anterior commissure. They extend beyond the margin of the cords making them uneven. *Acoustic finding:* Hoarse, occasionally bitonal cry. Tape recordings made before (A) and after (B) aspiration of the thrush colonies. **A**. *Sonagram No. 72a:* 0.7 to 2.1 s: Harmonic structure masked by noise, up to 1,3 s in the time range of the cry. A relatively well-defined, irregular, linelike component showing 2000 to 3000-Hz oscillations appears within the cry; it is presumably produced by the thrush colonies extending over the vocal cord margins. Excessive superposition of noise, especially up to 6000 Hz. A particularly intensive component appears at 2500 Hz. Due to the noise mask, the cry is strongly hoarse. A bitonal effect is caused by the simultaneous appearance of two kinds of components (the harmonic structure and the linelike component described above). **B**. *Sonagram No. 72b:* Regular cry with a characteristic harmonic structure. At sites (0.5–0.65 s), however, a double harmonic structure appears due to the remaining thrush colonies. No substantial masking by noise, but the individual overtones are frayed up to about 3500 Hz. Regular cry, slightly veiled.

the most common. Sound production is affected in the same way as in fibrinous laryngitis as the local changes are similar. There is hoarseness (Figures 7-33A), the severity of which may depend on the duration of the intubation and mostly of the laryngeal findings. It disappears within a few days when there is spontaneous remission (Figure 7-33B). In severe cases, after long intubation, necrosis of the vocal cords and of the cricoid cartilage may also occur (Figures 7-34A and B).

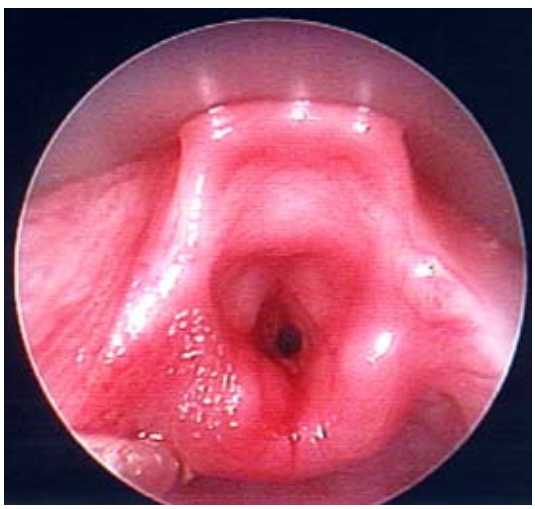


FIGURE 7-31. Postintubation glottic edema.

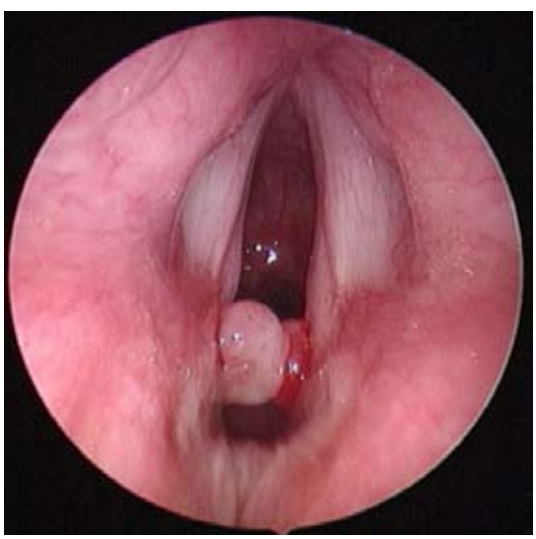


FIGURE 7-32. Postintubation granulomas.

Posterior glottic stenosis (PGS) is a disabling disease commonly induced by endolaryngeal injury from intubation or surgery (Roh, 2005). In the data of Rutter and Cotton, 2004) 29 patients raging in the age from 2 to 8 years were treated with isolated posterior glottic stenosis, among them 21 with a history of prolonged intubation. Endoscopic posterior cricoid split and rib graft insertion in children appears to be safe and effective in the management (Inglis, Perkins, Manning, & Mouzakes, 2003). Isolated PGS in children also is effectively managed with costal cartilage grafting of the posterior cricoid (Rutter & Cotton, 2004). We managed such a case with posterior snap-graft from rib cartilage (Figures 7-35A through E). The study of Roh (2005) suggests that topical mitomycin C can be helpful in preventing the progression of PGS.

Subglottic edema is another common complication of prolonged intubation (Figure 7-36); it is associated with a barking or hollow cough and a stridor of subglottic character (see the vocal changes in subglottic laryngitis, Figures 7-80 and 7-81.)

Subglottic cicatrization and stenosis are the most important late complications which often make a larynx widened operation necessary (The possible surgical methods which may be applied in these cases, are detailed later in this chapter.). Massive scar is not recommended to be treated by laser as an increase of the obstruction may be the final consequence (Figures 7-37 and 7-38). Inspiratory stridorphonation or stridor of subglottic

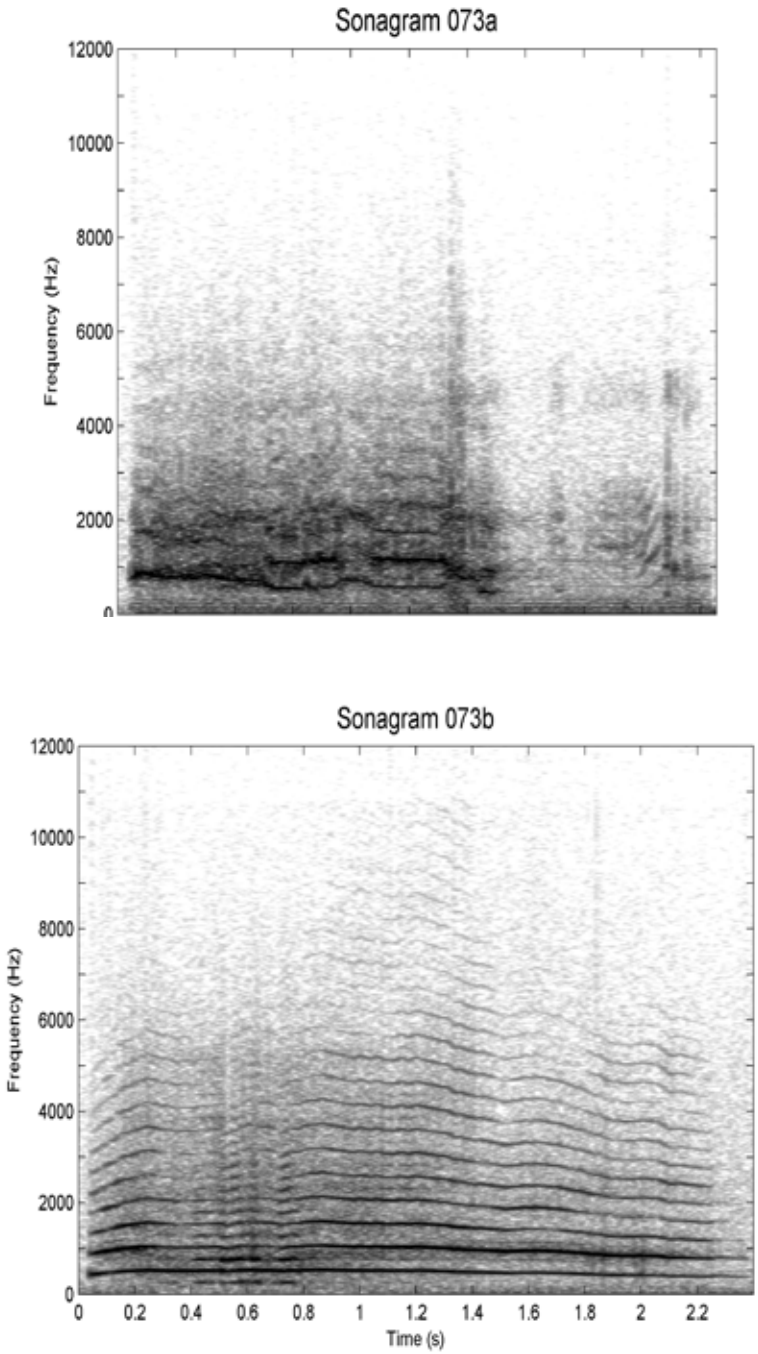
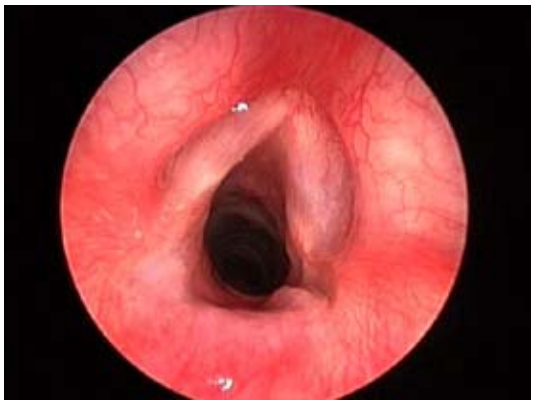


FIGURE 7-33. State after prolonged intubation. Child aged two and a half years. *History:* Several attempts at resuscitation and prolonged intubation (8 days) in shock and cardiovascular failure following abdominal operation. Tape recordings made 2 days (A) and 12 days (B) after extubation. *Laryngoscopy:* Exulceration, granuloma on both vocal cords. *Acoustic finding:* Extremely severe hoarseness, relieved spontaneously. **A**. *Sonagram No. 73a:* Sporadically appearing regular acoustic structure. Roughly normal overtones. Excessive masking by noise. Very hoarse cry. **B**. *Sonagram No. 73b:* Some of the overtones are extinguished in a part of the record (0.2–0.6 s), but there is a tendency toward assuming normal acoustic structure; essentially it is a regular crying sound.



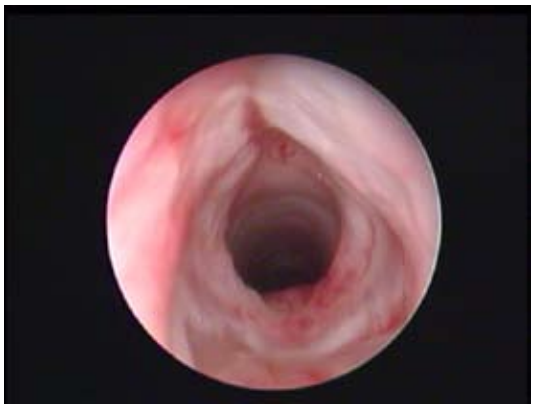
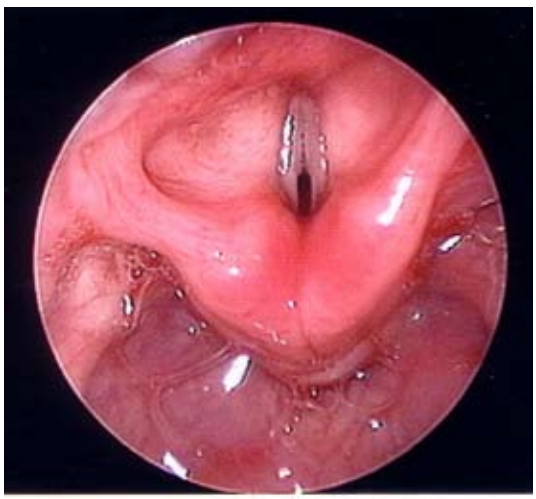
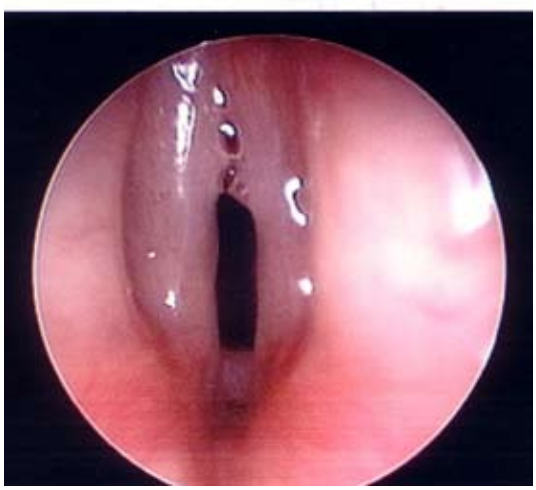
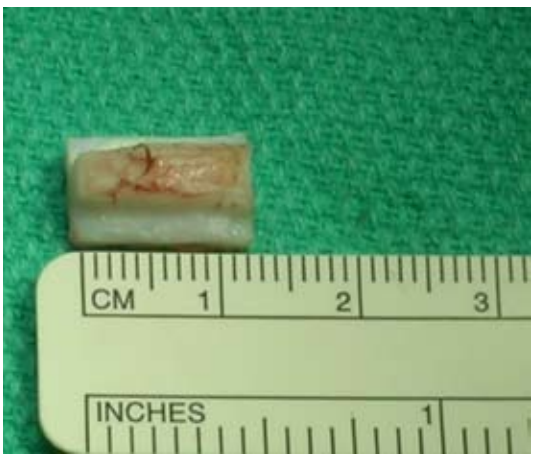


FIGURE 7-34. A. Postintubation necrosis of right vocal cord. **B**. Postintubation cricoid cartilage necrosis.



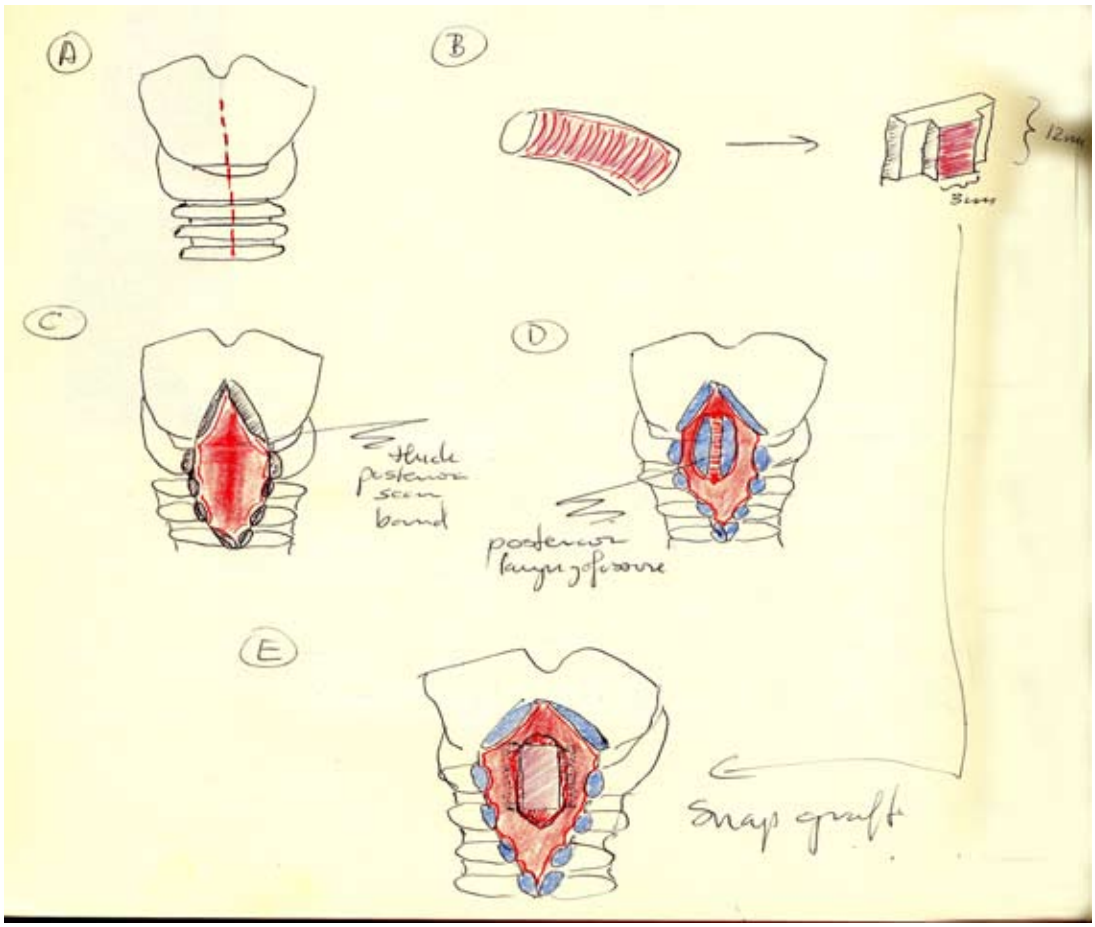


A

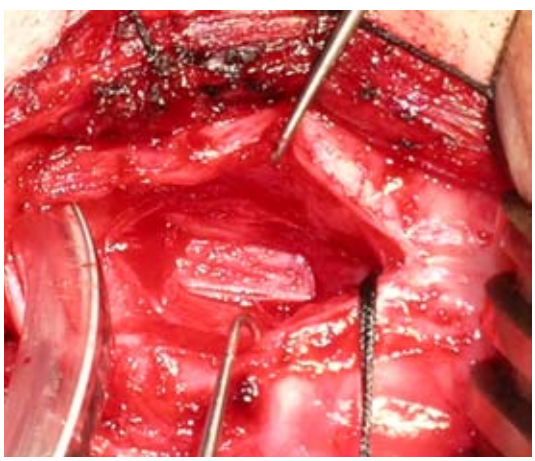


B

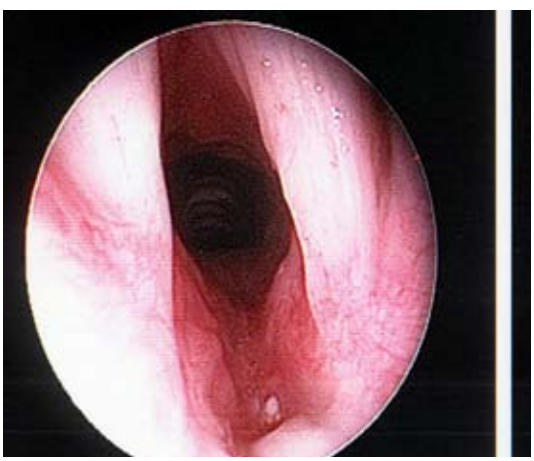
FIGURE 7-35. **A**. Posterior glottic stenosis in a 4-year-old child; **B**. posterior snap-graft from rib cartilage; **C**. Rib graft snapped into posterior cricoid split; **D**. 3 months after surgery; **E**. Koltai's drawing of the case.


A
B
12mm
C
Muscle
posterior
seam
band
D
posterior
laryngofissure
E
Snap graft

C



D



E

character, which may be associated with barking cough, are acoustic concomitants of the late complications involved the subglottis (Figure 7-39).

It is worth mentioning that acquired subglottic stenosis (similarly to posterior glottic stenosis, discussed above) may occur also as a consequence of gastro-esophageal reflux (Silva, 2000). In this case the initial evaluation should consist of a complete history and physical examination, flexible nasolaryngoscopy, barium esophagogram, and airway and chest radiographs. If the clinician has a high level of suspicion for reflux, then pH-probe and/or nuclear scintiscan should be done, and according to the studies, correct management introduced.

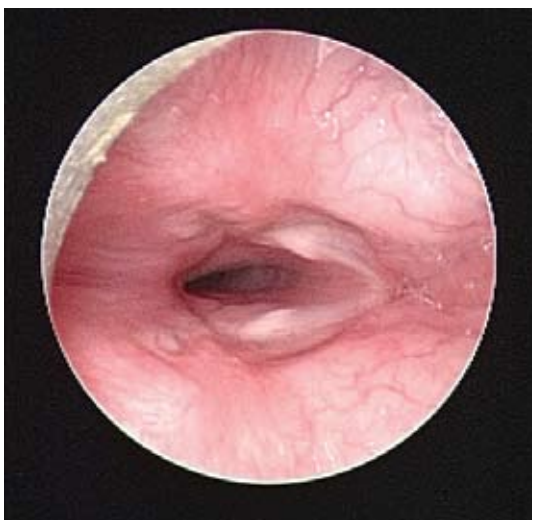


FIGURE 7-36. Subglottic edema after intubation.

KEY POINT

The early complications of a prolonged intubation are: hematoma, edema, exulceration, granulation, and—in severe cases—necrosis of the vocal cords which cause hoarseness. Subglottic edema is another consequence with hollow or barking cough and stridor. The most important late complication is subglottic stenosis with stridor of subglottic character and with hollow cough. Laryngoscopy is in all cases indicated. Massive and permanent glottic and subglottic cicatrization often needs larynx -surgery (see later section).

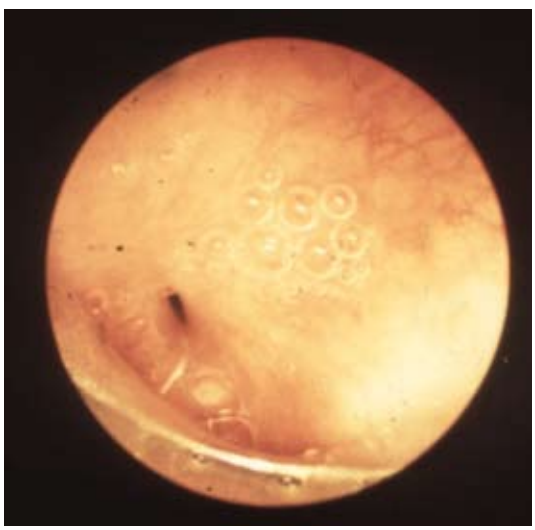


FIGURE 7-37. Severe subglottic stenosis after prolonged intubation (lasting 1 month), and after repeated laser management in a child aged one and a half years.

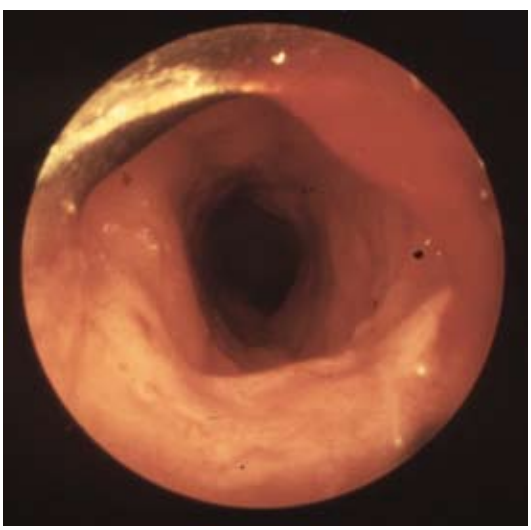


FIGURE 7-38. The same child (see Figure 7-37) after reconstruction surgery. See the placed oral mucosa in the posterior laryngeal wall.

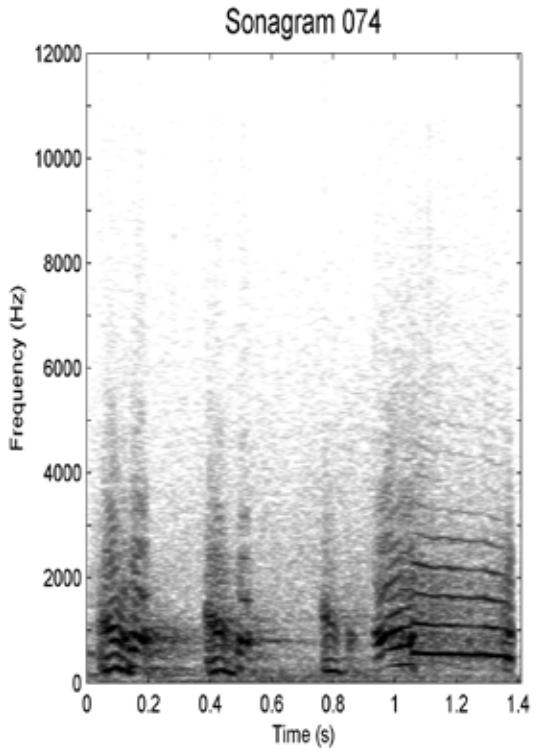


FIGURE 7-39. Condition after prolonged intubation. Subglottic stenosis. Infant aged 6 months. *History:* Convulsions, intubation because of respiratory distress at the age of 5 days. Prolonged intubation for 15 days. Satisfactory condition after 2 months' treatment in hospital. Later occasional episodes of "malaise" and cyanosis. *Laryngoscopy:* Laryngeal inlet normal, good mobility of vocal cords. In the subglottis, circular, scarred stenosis, visible mainly laterally and posteriorly. *Acoustic finding:* Stridor-phonation and hollow cough. *Sonagram No. 74:* Three consecutive coughing sounds, then phonation-like stridor (0.9–1.4 s). In the coughing phase the dense noise band is arranged overtonelike, whereas in the stridorous phase the overtones are 600 to 700 Hz apart.

Papilloma of the Larynx (upper respiratory papillomatosis)

Papilloma of the larynx is rare in infants, whereas *it is the most common laryngeal tumor in young children* (Lábas, 1968b), and the second most frequent cause of childhood hoarseness (Shehab, Sweet, & Hogikyan, 2005). The primary growth is usually located on the vocal cords (Figures 7-40 and 7-41) spreading from there to the deeper airways, into the trachea and bronchi or upward, to the pharynx, entry of esophagus, palate (Figure 7-42), uvula and nose. Extralaryngeal spread of respiratory papillomas has been identified in approximately 30% of children (Derkay, 1995). Soldatski, Onufrieva, Steklov, and Schepin (2005) treated 448 children with recurrent respiratory papillomatosis in St. Vladimir Moscow Children's Hospital between 1988 and 2003, among them papillomas extension down to lower airways was observed in 40 children (8.9%). Consultant members of the British Association of Paediatric Otorhinolaryngology (Tasca, McCormick, & Clarke, 2006) sent around a questionnaire on current practice concerning various data and management

of recurrent respiratory papillomatosis (RRP) in the paediatric population in the United Kingdom. From the results: distal spread of RRP has occurred in 27 (26.2%) children among the total 103 patients.

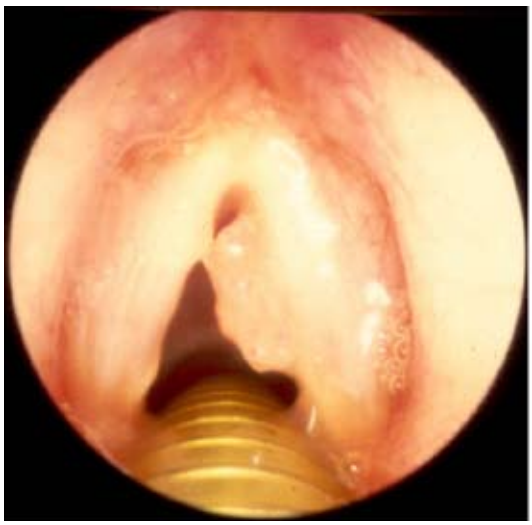


FIGURE 7-40. Papilloma on the right vocal cord in a child aged 1 year.

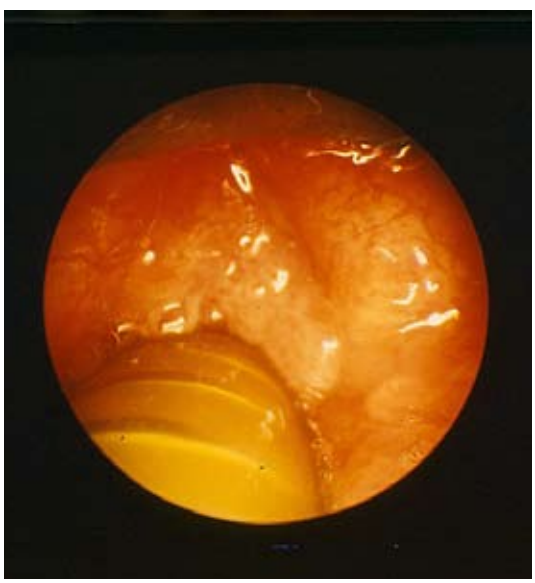


FIGURE 7-41. Laryngeal papilloma covered mostly the whole glottic lumen in a child aged 2 years. Very hoarse, sometimes aphonic cry.

Because the papillomatosis may not be confined to the larynx and can be seen in any portion of the upper aerodigestive tract, the term recurrent respiratory papillomatosis (RRP) has been proposed for the disease entity by Myer and Cotton (1988). The growths are berrylike structures, dirty pink in color.

The symptoms depend on the size and location of the tumor. There is more or less severe impairment of the two laryngeal functions in the case of a papilloma in the larynx: respiration and phonation, and thus the major symptoms are stridor, dyspnea and—in extreme cases—apnea, as well as hoarseness or aphonic cry. Of the two kinds of symptoms the voice disorder is the more characteristic.

The cry may exhibit changes other than hoarseness: it *may be bitonal, wan, or shrill* (Figure 7-43). The acoustic structure is highly unstable. The harmonic structure is haphazard: the number of harmonics varies widely, they may suddenly appear or disappear. This can be attributed to the furrowed, uneven surface of the tumor, and to the fact that papilloma in infants

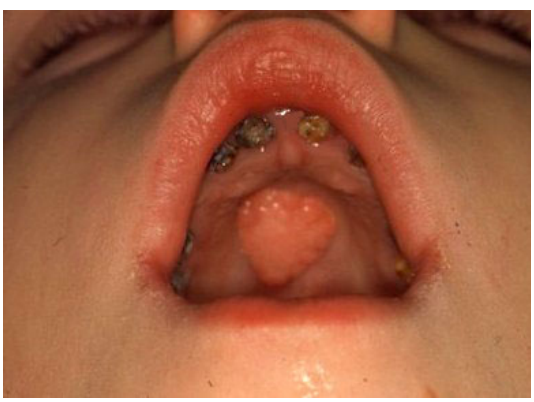


FIGURE 7-42. Papilloma of the palate.

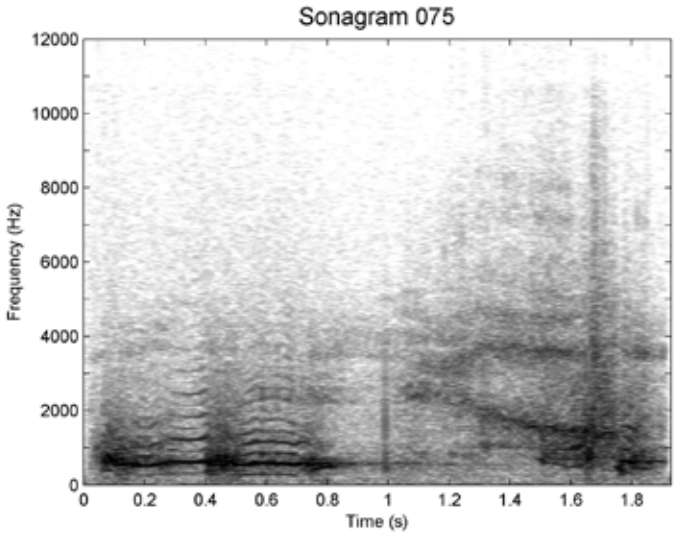


FIGURE 7-43. Laryngeal papilloma. Child aged two and a half years. *History:* Hoarseness noted by the parents for about a year. *Laryngoscopy:* On both vocal cords and in the anterior commissure raspberrylike, grayish-pink structures. The size of the entire mass is about that of half a bean. *Acoustic finding:* Hoarseness. *Sonagram No. 75:* 1.2 to 1.9 s: Mutilated harmonic structure, in which the formant structure of the sound [a:] (long *a* as in *far*) can be recognized vaguely in the Hungarian word *jaj* (= *ouch*). The overtones are replaced by thick bands of noise; no other noise components are visible. Verbalization has no major significance as regards the character and acoustic assessment of the sound.

and children—unlike in adults—is much more often multiple than solitary.

Extralaryngeal location may alter the symptoms, for example, tracheal stridor or cough; in other patients nasal secretion and occasionally hemorrhage also may develop.

Diagnosis can be made by laryngoscopy (rigid airway endoscopy, microlaryngoscopy, eventually associated with bronchoscopy in cases suspected to tracheal invasion and sometimes nasopharyngoscopy, if the lesion occurs in the nose or on the posterior wall of the palate), and histologic examination.

Vocalization improves upon removal of the papillomatous growth, but it usually fails to clear up completely owing to residual changes (epithelial lesion, uneven surfaces).

As for the management, no single modality has been effective consistently in eradicating RRP (Derkay & Darrow, 2000). *The current standard is surgical therapy* with a goal of maintaining the free airway and doing no harm. Cup forceps or CO_2 laser may be used; the latter has been favored over cold instruments in the treatment of RRP involving the larynx, pharynx, upper trachea, and nasal and oral cavities. Potential complications of laser surgery include the development of anterior commissure webs, laryngeal and subglottic stenosis, and the possibility of a laser-induced endotracheal tube fire. At Cincinnati Children's Hospital, 21% of patients required a long-term tracheostomy (Cole, Myer, & Cotton, 1989). Decannulation should be considered as soon as the disease is managed effectively with endoscopic tech-

niques. According to Huber, Sadick, and Gotte (2005) microdebrider and CO_2-laser are currently the most widely used surgical options.

At present, the most commonly recommended adjuvant medical therapy is based on cidovir and alpha-interferon. Children requiring laser therapy more frequently than 6 times in 12 months or who have evidence of distal spread of RRP should be considered for adjuvant interferon therapy (Derkay & Darrow, 2000). A maximal effectiveness of alpha-interferon therapy was revealed in RRP patients with human papillomavirus (HPV) type 6 as compared with HPV 11 (Gerein V., Rastorguev, Gerein J., Jecker, & Pfister, 2005). This observation indicates the necessity of HPV typing. Side effects also should be monitored. According to our findings in Heim Pál Children's Hospital, Budapest, children with interferon treatment showed a decrease in the growth rate of papillomas and the time between two surgical interventions increased (Hirschberg, 1989). The American Society of Pediatric Otolaryngology members (Schraff et al., 2004) experienced an evolution in the past decade toward the increased use of antiviral adjuvant therapy (cidovir and interferon) and the use of microdebrider techniques for surgical management. Cidovir may be administered intralesional or intravenous. According to many up-to-date publications (Dikkers, 2006; Mandell et al., 2004a; Pontes, Avelino, Pignatari, & Weckx, 2006; Pudszuhn, Welzel, Bloching, & Neumann, 2007; Sheahan, Sexton, & Russell, 2006) intralesional injection of cidovir (dose of 5 mg/1mL) is an adjuvant, a promising and powerful therapeutic approach but not a curative therapy. The greatest beneficial effect was seen after the fourth injection. Successful outcomes also have been reported with intravenous cidofovir, though on the basis of a limited number of instances (Sheahan et al., 2006). Currently, the combination of microsurgery with intralesional cidofovir seems to be the treatment of choice for RRP (Dikkers, 2006). Recently, Shikowitz et al. (2005) reported on advantageous efficacy with the use of photodynamic therapy with meso-tetra (hydroxyphenyl) chlorine photosensitizer which may reduce the severity of respiratory papillomatosis possibly through an improved immune response. Recent advances in immunologic research offer the hope of immune system modulation as potential future treatment modalities (Huber et al., 2005).

KEY POINT

Papilloma of the larynx is mostly located on the vocal cords which may cause –depending on its measures—hoarseness, aphonia, sometimes also bitonal, wan or shrill cry with stridor and dyspnea. The papilloma often spreads upward: to the pharynx, entry of esophagus, palate, and nose, and to the deeper airways (trachea and bronchi). The symptoms depend on the localization. The current standard in therapy is surgery, alpha-interferon, and cidovir.

In most of the children remission and spontaneous improving may be anticipated around puberty; however, in cases extending over the adolescence or adulthood, in a few percent of cases malignization can occur. The prognosis for the disease after development of pulmonary papillomatosis is always serious (Soldatski et al., 2005).

The combination of clinical and characteristic radiologic features (chest radiograph, CTI) suggests a diagnosis of pulmonary spread of laryngeal papillomatosis (Abe et al., 2006). In the case of a tracheal and bronchial spreading one should consider a malignant degeneration.

Case Study

History of a patient with respiratory papillomatosis. The female patient was 1 year old when the first symptoms occurred: impeded nasal breathing and diffuse nasal secretion. Adenotomy was performed in the ENT Department of the County Hospital in Székesfehérvár. During the surgery it was established that the adenoid was clutched with papillomalike tissue. The infant was then sent to us, the Ped. ORL Department of the Heim Pál Hospital for Sick Children, Budapest. The diagnosis: papilloma was histologically verified. In the next 3 years repeated excision of papillomata from the nasopharynx, soft palate, tonsillar pillars, nose, entry of esophagus, and larynx (vocal cords); the latter was accompanied with severe hoarseness. At age 4 years a cardiosurgical intervention was performed because of congenital heart anomaly in a New Jersey hospital, USA. Two days before the flight we removed papilloma tissue from the trachea. After 6 symptomless years renewed occurrence of the papilloma in the palate and later again in the nose occurred with repeated surgery. The suggested interferon therapy was contraindicated by the cardiologists. The nasal surgery was performed with traditional instruments, later by laser in another public institute with the consequence of severe narrowing of one nasal aperture and large perforation of the septum. The patient was 18 years old when a cough developed not influenced by routine management. Bronchoscopy: papilloma in the trachea (Bronchological Department of the County Hospital in Székesfehérvár), repeated cryosurgeries. The histologic examination did not reveal any malignancy during the past 15 years. The cough did not cease after removing the tracheal papilloma and hemoptoe occurred. MRI and CT examination found a tumor in the sixth and tenth segment of the caudal lobe of the left lung; therefore, the concerning lobe was removed (Chest Surgical Department of the Korányi Lung Institute, Budapest). Malignant deterioration could not be justified histologically: HPV pulmonis was verified. At present, interferon and cidovir therapy are planned, after reconvalescence is complete.

Vocal Cord Polyp, Laryngeal Fibroma

Polyp and fibroma occur less commonly in the larynx of infants and young children than papilloma (Romanet, Thuel, Duvillard, Schweighoffer, & Chevrier, 1997). Polyps usually originate on the vocal cords or occasionally on the false vocal cords. The etiology is not known, although its development has been connected with overexertion and abusus of the voice in hyperactive, impulsive children aged mostly 5 to 6 years and may be considered a sequela of a phonotrauma. In infants and toddlers it may be due to much crying. According to Chalabreysse, Perouse, Cornut, Bouchayer, and Loire (1999) congenital benign vocal cord lesions are certainly more common than is usually thought. Vocal cord polyp is a heterogeneous group of benign lesions. Coulombeau, Perouse, Chalabreysse, and

Faure (2006) regard it as an inflammatory false tumor of the larynx.

Polyps are baglike edematous, myxomatous or teleangiectatic bulges on the vocal cords; they are pseudotumors with smooth, glassy, or globular surface and with gray, yellowish or dark-red color (Wendler, Seidner, & Eysholdt, 2005). Histologically, long-term vocal polyps have thin or discrete thickening of basement membrane, pronounced edema, and dilated vessels. *Fibroma is a dirty white structure* and always less multiple than papilloma. These false tumors have to be differentiated from the laryngeal neurofiboma (in connection with or not related to Recklinghausen's disease) which can also occur in children (Chen, Lee, Yang, & Chang, 2002; Fukuda, Ogasawara, Kumoi, Sugihara, & Wada, 1987; Soboczynski, Woznicki, & Sobala, 1989).

Diagnosis of polyps and fibromas is made by laryngoscopy, if the change in phonation excites suspicion. Management: excision with the aid of microlaryngoscopy.

Hoarseness is the main symptom. It is usually marked, but may vary in severity, and may be associated with a slightly pressed cry (Figure 7-44).

KEY POINT

Polyp and fibroma of the vocal cord is consequence of vocal abuses and may be connected with much crying, causing hoarseness and pressed phonation. Excision by means of microlaryngoscopy is recommended.

Laryngeal Cysts

Laryngeal cysts are uncommon congenital anomalies of the larynx. The two types of cysts are: saccular due to obstruction of the laryngeal saccule orifice in the ventricle leading to retention of mucus and ductal which arise from blockage of submucosal mucus glands (Tewfik & Sobol, 2003). The congenital cysts are almost without exception retention-type in origin due to obstruction of the orifice of small mucous glands (Lellei, 2004). *Laryngeal cysts may develop in several parts of the region.*

The most frequent localization is the vallecular cyst (Figures 7-45A, B, and C). It develops from the false vocal cord and may spread to the glottis or the hypopharynx, sometimes causing significant obstruction of the airways. In these cases intense inspiratory stridor may occur (Tuncer et al., 2002), associated with feeding difficulties (Hsieh et al., 2000). The crying sound is generally clear. According to Yao, Chiu, Wu, and Huang (2004) vallecular cyst is a rare but dangerous cause of stridor in neonates and young infants. According to the authors the vallecular cyst may be associated with laryngomalacia and gastroesophageal reflux. *Vallecular cyst may cause severe upper airway obstruction*; thus, the antenatal diagnosis with ultrasound and a planed, careful perinatal management is very important as recommended by Cuillier, Samperiz, Testud, and Fossati (2002), on the basis of a case diagnosed at 28 weeks of gestation.

As an extremely rare case, Lee et al. (2000) found *epiglottic cyst* 7 hours after birth, causing airway obstruction and *inspiratory stridor.*

Intracordal cyst is also a curiosity. Smith et al. (2000) were the first to present a record and a photograph of a case occurring in a neonate with symptoms of *episodic*

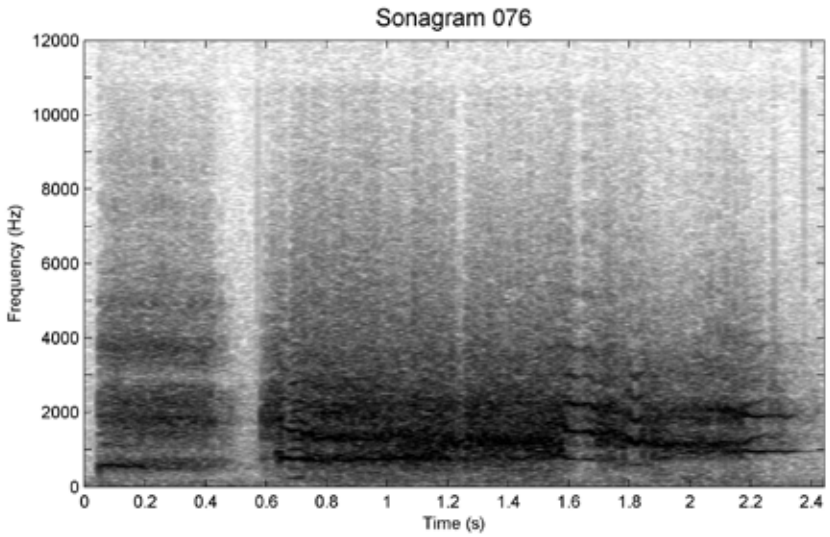


FIGURE 7-44. Laryngeal fibroma. Child aged two and a half years. *History:* Hoarseness from birth. *Laryngoscopy:* Bean-sized, whitish-pink structure. *Acoustic finding:* Hoarseness. *Sonagram No. 76:* Fundamentally normal, but less distinct sound components. The acoustic structure is strongly masked by noise due presumably not only to hoarseness, but also to pressing. This proportion of normal acoustic structure and noise produces a very hoarse cry. A fundamental frequency runs like a line trough the frequency range of 700 to 800 Hz. The higher overtones show

stridor, respiratory distress, and feeding difficulties.

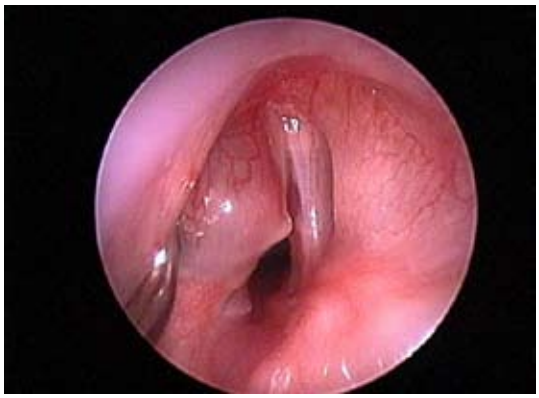
The subglottic form is also infrequent. Agada, Bell, and Knight (2006) observed seven patients with pediatric subglottic cysts, and stridor during 10 years (1995–2005) at the Leeds teaching hospitals: five were born premature and all were intubated with an average intubation period of 20.6 days. Six of their patients underwent endoscopic marsipulization using cup forceps and one with a contact diode laser. On the other side, according to Watson, Malik, Khan, Sheehan, and Rothera (2007) the number of cases with acquired paediatric subglottic cyst has been increasing over the last 3 decades which represent the fourth most common causes of airway obstruction in their series. Figure 7-46 shows our own case which caused severe respiratory problems: *stridor and barking cough*. In the case of Bokhari et al. (2004) the subglottic cyst was associated with complete tracheal ring deformity which occluded the airway completely.

Recently, Stewart, Cochran, Iglesia, Speights, and Ruff (2002) published a case with *tracheal bronchogenic cyst*: as an unusual cause of stridor and wheeze of an infant.

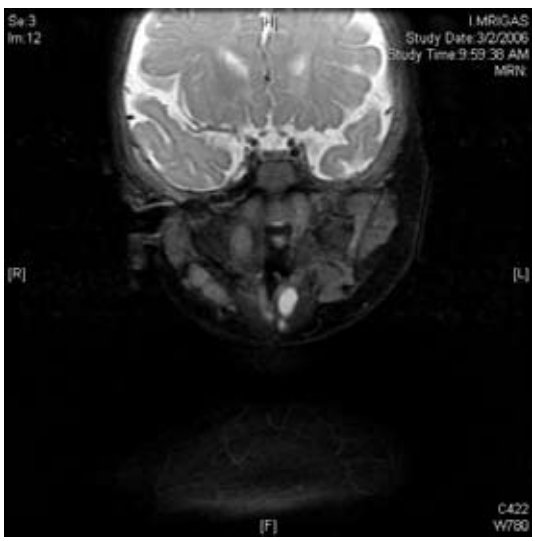
Diagnosis can be made by endoscopy supplemented with MRI (see Figure 7-45B) or with ultrasound investigation (Lellei, 2004).

Regarding management, needle aspiration or incision of the lesion and aspiration of the mucus result in rapid improvement (Figures 7-47A and B).

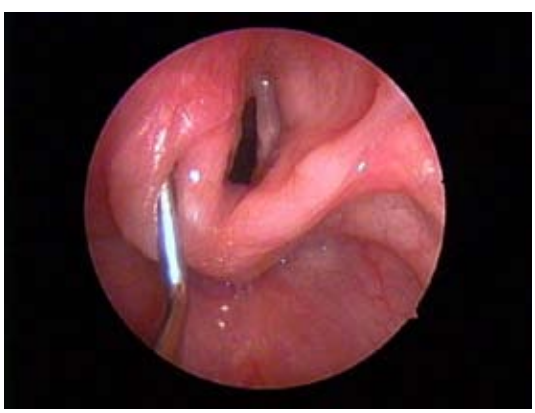
If the cyst becomes repeatedly full with mucus marsupialization should be performed (see Figure 7-45B). Very rare external opening and a total removal of



A



B



C

FIGURE 7-45. A. Saccular cyst of a newborn; **B**. MRI picture; **C**. 1-year postop.

the cyst is necessary. A new system of classifying for congenital laryngeal cysts was proposed by Forte, Fuoco, and James (2004) based on the extent of the cyst and the embryologic tissue. This classification can help guide the surgeon. Type I cysts are confined to the larynx, the cyst wall being composed of endodermal elements only, and can be managed endoscopically. Type II cysts extend over the confines of the larynx containing endodermal and also mesodermal elements (epithelium and cartilage) in the wall and require an external approach.

KEY POINT

The laryngeal cyst may develop in several parts of the region. The most frequent localization is the vallecula, which causes severe respiratory problems with stridor. It may occur on the epiglottis with inspiratory stridor, intracordal with hoarseness, and subglottic with deep stridor and barking cough. If sucking of the fluid (punction) and incision is not enough in the management, marsupialization is suggested.

Laryngocele

Laryngoceles are air-filled dilatations in the laryngeal saccule that communicate with the laryngeal ventricle. Internal laryngoceles arise in the anterior ventricle and extend posteriorly and superiorly up the false vocal cord and aryepiglottic fold, but takes place entirely inside the laryngeal cartilage. External laryngoceles extend cephalad through the thyrohyoid membrane (Hughes & Dunham, 2000). The external laryngocele could be touched and seen on the neck, which may enlarge during coughing. The internal one resem-

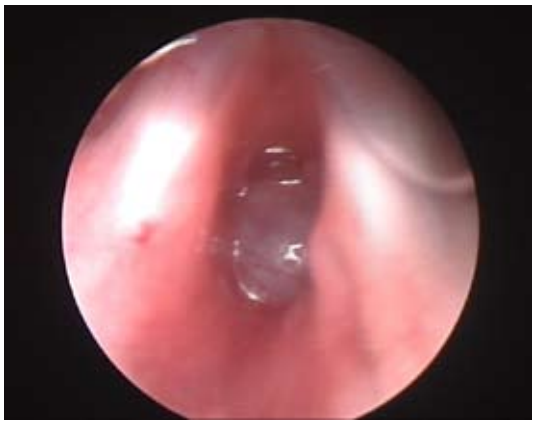


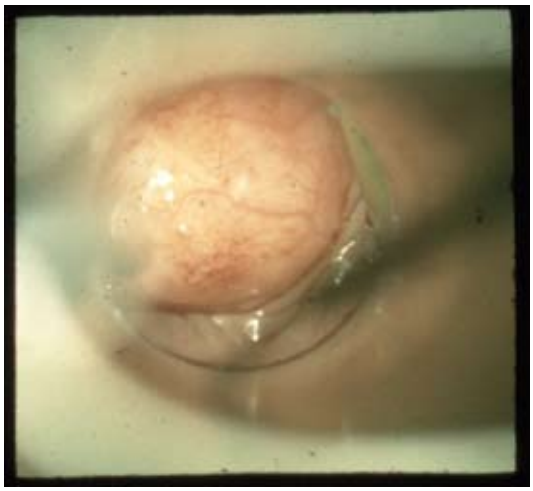
FIGURE 7-46. Subglottic cyst in a child aged 14 months.

bles a laryngeal cyst. The most important separating criterion between cyst and cele according to us (Hirschberg, 1989) is that the laryngoceles are fulfilled with air and communicate with the laryngeal lumen, in contrast with cysts which are closed and their lumen is full of fluid. The laryngocele may infected secondarily and then a pyocele develops.

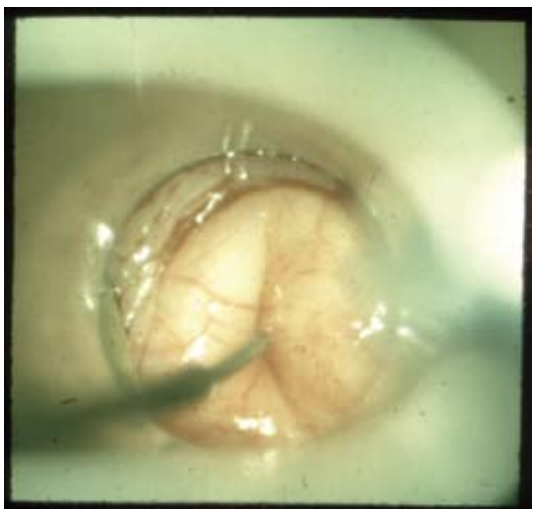
KEY POINT

Laryngocele is an air-filled dilatation of the laryngeal saccule and communicates with the laryngeal lumen. It may be infected secondarily and then a pyocele develops. In infants and young children it is a rare anomaly. Large celes cause inspiratory stridor. Its diagnosis can be made with endoscopy and MRI. The management is surgical.

A little laryngocele does not generally cause complaints. *In the case of a larger one*, especially if it is fulfilled with fluid, an *inspiratory stridor may be heard*. It is sometimes intermittent and more severe when the child lies on the side opposite to the cele. Dyspnea is rare. The cry is clear.



A



B

FIGURE 7-47. A. Vallecular cyst in a child aged 2 years. **B**. The same child during the sucking of the fluid.

As for diagnosis, endoscopy and imaging methods: plain x-ray films of the neck, ultrasound, computed tomography (CT), MRI are helpful (Chu, Gussack, Orr, & Hood, 1994) as demonstrated in our own case (Figure 7-48).

The management is surgical through the mouth or with an external opening method (Figure 7-49). The choice of the proper

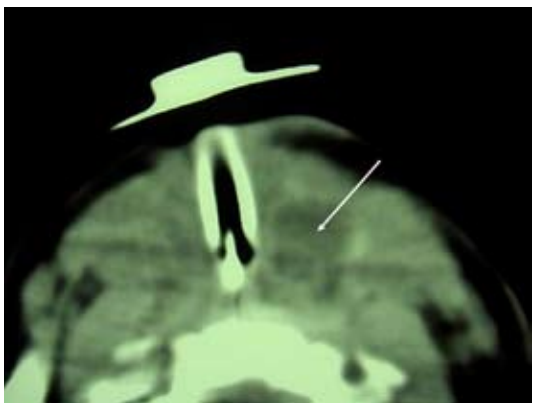


FIGURE 7-48. MRI of a laryngocele in a child aged one and a half years.

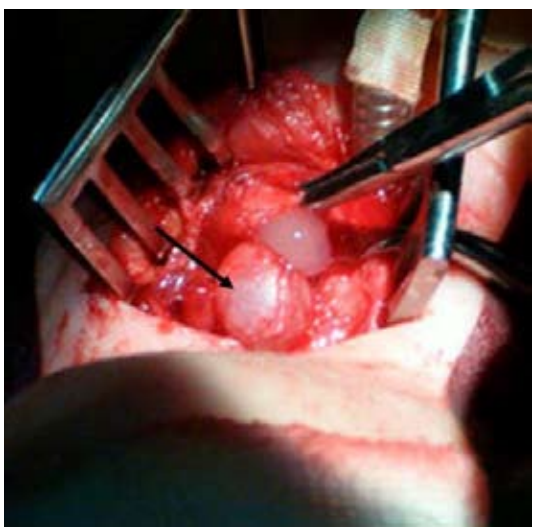


FIGURE 7-49. Same child during surgery.

depends on the situation of the individual child. A surgery in infancy and young children is infrequent as opposed to respective cases in adults (Pirsig, 1980).

Laryngeal Cleft

The laryngo-tracheo-esophageal cleft was first described by Richter in 1792, and in modern times Finlay (1949) discovered it again. This condition *results from the failure of fusion of the posterior cricoid lamina during gestation*. The first successful operative reconstruction was performed by Peterson (cit: Narcy et al., 1979). Until the 1980s some 80 cases were published in the literature. Recently Kubba, Gibson, Bailey, and Hartley (2005) presented an update to the Great Ormond Street Hospital (London) series of laryngeal clefts, describing 35 further instances of clefts between 1992 and 2003. Rahbar et al. (2006) published a 10-year retrospective study (1994–2004) about 22 patients (mean age: 21 months) with laryngeal cleft in two pediatric tertiary care medical centers.

Several classifications of this anomaly are known. The most practical one is the arrangement along the lines of the extension of the cleft. The most mild form refers only to the cricoid cartilage, the other one expands to the subglottic area and the superior part of the trachea, the most severe form reaches until the carina longwise of the entire trachea. The cleft develops around the 35th day of the gestation period due to the disorder of rostral development of the tracheoesophageal septum, which hinders the fusion of the posterior part of the cricoid cartilage.

The diagnosis of this uncommon anomaly is not always easy, as the radiologic symptoms are not significant and the endoscopic picture also may be very poor (Holinger, Tansek, & Tucker, 1985). Condon, Salvage, and Stafford (2003) recommend CT scan in evaluation. Endoscopy, however, is unavoidable in diagnosis.

Frequent aspiration, respiratory distress and coughing (especially during feeding), repeated pneumonia, and backwardness in augmentation of weight dominate the clini-

cal picture. Mortality is high as diagnosis is mostly not available in due time.

KEY POINT

The laryngeal clefts may be classified in terms of their extension. The most common and mildest form touches on only the cricoid cartilage whereas the severe forms reaches the entire trachea. Frequent aspiration, repeated pneumonia, and cough are the most typical symptoms. Diagnosis (also with endoscopy) may be difficult. The management is surgical.

It is very important to prevent the occurrence of these severe symptoms; therefore, first the application of a gastric tube and surgery as early as possible is indicated (Watters & Russell, 2003). Each patient should be assessed properly, and the surgical approach should be individualized based on the symptoms, other associated findings on airway endoscopy, and type of cleft (Rahbar et al., 2006). Koltai, Morgan, and Evans (1991) published an article about endoscopic repair of supraglottic laryngeal clefts. Associated congenital anomalies are common. Mitchell, Koltai, Matthew, Bailey, and Evans (1989) reported 3 cases of severe tracheobronchomalacia associated with laryngeal cleft: each of the children had a major cleft, type 3 in Evans' classification. Each of these conditions is difficult to treat separately. In combination these major airway anomalies cause management problems of great complexity, too. Most type 1 and smaller type 2 clefts may be repaired endoscopically whereas larger clefts should be repaired through an anterior approach (Kubba et al., 2005). Type 4 laryngotracheoesophageal clefts are best repaired via an anterior cervicothoracic approach, with or without a median sternotomy, and extracorporeal membrane oxygenization (Mathur, Peek, Bailey, & Elliot, 2006). *Early diagnosis and surgical repair are essential for successful treatment.*

Laryngomalacia (chondromalacia laryngis; soft, flaccid larynx)

This is the most common cause of laryngeal stridor (Archer, 1992; Merrot, Fayoux, Vachin, Chevalier, & Desaulty, 2004; Midulla et al., 2004; Tewfik & Sobol, 2003), that is, the classical form of congenital inspiratory stridor described in pediatric textbooks (Fanconi & Wallgren, 1972; Nelson, 1969). Zalzal (1989) found 59.8% laryngomalacia in cases with laryngeal anomalies. In our statistics (Hirschberg, 1989) this anomaly is represented 31% among the organic laryngeal alteration of infants and young children. Male infants are affected twice as often as female (Tewfik & Sobol, 2003). The essence of this anomaly is the congenital weakness of the cartilaginous laryngeal structure, and flabby, flaccid laryngeal mucosa (Cotton & Reilly, 1983), often associated with omega-shaped epiglottis, and sometimes with shortening of the aryepiglottic folds (Figure 7-50 and 7-51).

A variety of mechanisms have been reported to explain the etiology, the causing factors, including cartilage immaturity, and poor neuromuscular control secondary to hypomaturity or dysfunction (Archer, 1992). Lately, the possibility of gastroesophageal reflux (GER) has been raised in the etiology. According to Tewfik and Sobol (2003) histologic evidence of gastroesophageal reflux laryngitis recently has been documented from aryepiglottic specimens. This idea, however, is not ac-

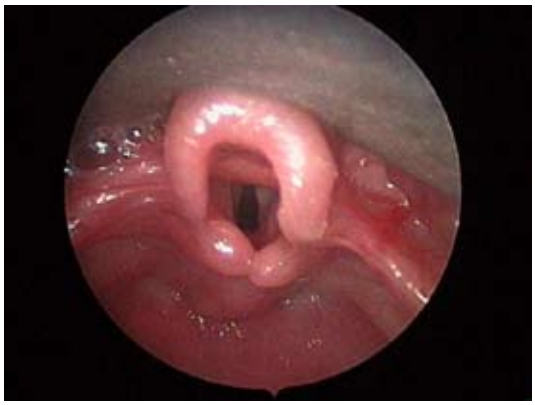


FIGURE 7-50. Laryngomalacia; moderate to severe.

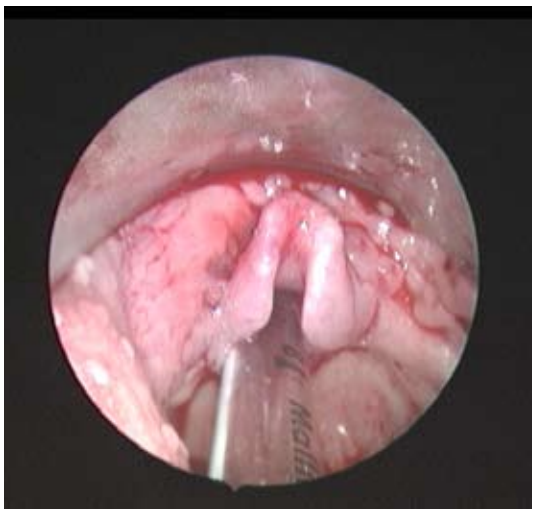


FIGURE 7-51. Laryngomalacia, severe, with epiglottic prolapse into the larynx.

cepted by some other authors (Giannoni et al., 1998), because after a 30-year history of GERD, at present there is no way to objectively measure the extraesophageal reflux disease (Czigner, 2002) and also according to Rosbe et al. (2003) it is difficult to determine the correlation between reflux and the upper airway symptoms. We agree with Stavroulaki (2006) in that at present limited evidence seems to exist to support a causative relationship between reflux and any otorhinolaryngologic condition or the effectiveness of treatment. In our opinion, this statement especially concerns laryngomalacia.

Inspiratory stridor in laryngomalacia is phonation-like (mostly noiseless or only slightly noisy), and, in addition, is usually cackling (Figure 7-52). Cackling stridor, noted for the undulation of overtones, can only be found in laryngomalacia. Its development can be explained by the state of the arytenoid cartilages, aryepiglottic folds, and loose, flabby laryngeal mucosa being sucked into the larynx on inspiration. This also can be demonstrated laryngoscopically. The stridor is worsened while crying or in an excited state. Prone position with the head up improves the stridor. Suprasternal retraction, and substernal retraction simultaneously with the stridor is a common sign. Also pectus excavatum may be seen occasionally and, according to some authors, as a consequence, but the relationship with laryngomalacia may remain unclear. *The cry of these infants is invariably a clear, regular cry.*

Stridor in laryngomalacia can be heard 2 to 3 weeks after birth. It will be louder due to increasing agility of the infant over the next weeks or months of life; respiration is then normalized in the age of 1½ to 2 years, as the connective tissue and cartilaginous structure of the larynx becomes stronger.

Midulla et al. (2004) report on episodes of microaspiration in 14 infants with laryngomalacia having been established with bronchoalveolar lavage. Severely affected children are at risk of feeding difficulties, apneic episodes and cor pulmonale secondary to upper airway obstruction (Martin, Howarth, Khodaei,

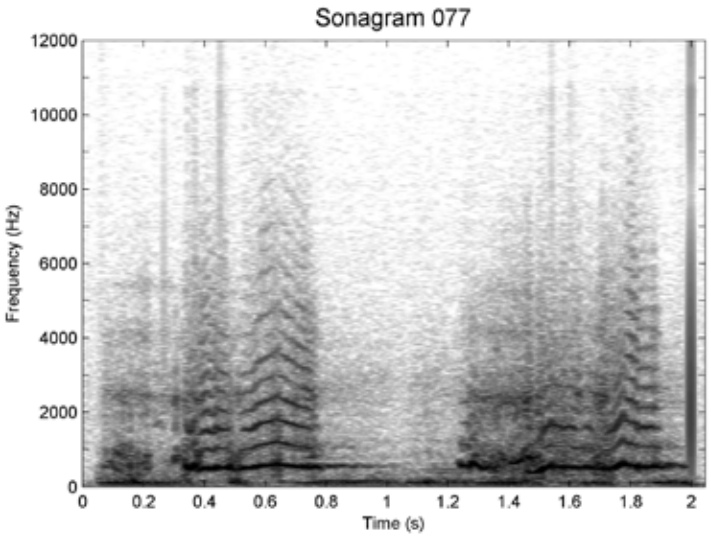


FIGURE 7-52. Laryngomalacia. Infant aged 12 months. *History:* Stridor from the age of a few weeks. *Laryngoscopy:* Intact vocal cords, good mobility. Flabby laryngeal mucosa, which is hypertrophic and lobular mainly in the vicinity of the right false vocal cord. *Acoustic finding:* Cackling stridor. *Sonagram No. 77:* Regular, but to some extent scanty structure of overtones; harmonics about 500 Hz apart with some hill-like elevations (0.8 and 1.8 to 1.9 s). At 2400, 4000, and 5400 Hz weak noise bands appear and some overtones are thickened. The rise and fall of overtone frequencies lend a cackling character to the stridor. Phonation-like stridor.

Karkanevatos, & Clarke, 2005). According to Vollrath (2004) life-threatening symptoms occur in 7 to 22% of all children with laryngomalacia, Merrot et al. (2004) indicate less favorable prognosis in 10 to 15% of cases.

The characteristic stridor occurs in laryngomalacia with a high degree of probability. *The diagnosis can be verified*, however, *only by laryngoscopy*. As stressed by Cotton and Reilly (1983): "Prior to endoscopy, there is no diagnosis, only impressions." According to Sivan, Ben-Ari, Soferman, and Derowe (2006) fiberoptic flexible laryngoscopy is the diagnostic procedure of choice in patients with laryngomalacia and the diagnosis is more accurate using anesthesia or sedation, whereas Yuen, Tan, and Balakrishnan (2006) prefer the rigid endoscopy in the evaluation of laryngomalacia which proved to be an appropriate and safe method in their practice. In any cases, direct laryngoscopy has to be performed with great care because introduction of the tube may alter the picture by extending the loose, cartilaginous larynx (the stridor, too, may transiently disappear or decrease). In severe forms, Valera, Tamashiro, de Araujo, Sander, and Kupper (2006) evaluated patients by means of a questionnaire given to the parents and with polysomnographic examination. In our opinion the latter also may help in the surgical indication. Audio-video recording is essential; this opinion is stressed also by Bent (2006).

Laryngomalacia in itself does not cause worry; severe respiratory symptoms occur seldom. In these cases various types of endoscopic laryngeal surgery are recommended and have been performed by us (Figures 7-53A, B, C, and D). Different formes of sur-

gery also are published in the literature: epiglottoplasty (Pucher & Grzegorowski, 2006), aryepiglottoplasty (Martin at al., 2005), epiglottopexy (Whymark, Clement, Kubba, & Geddes, 2006), laser resection of arytenoids mucosal excess associated, if necessary, with suprahyoid epiglottectomy (Merrot et al., 2004), CO_2 laser supraglottoplasty (Vollrath, 2004; Lee, Chen, Yang, & Chen, 2007). Toynton, Saunders, and Bailey (2001) report about 100, and Merrot et al. (2004) about 33 successful operations. In the database of the Heim Pál Children's Hospital, Budapest, laryngeal endoscopic surgery was performed in 10 patients, generally with conventional laryngomicrosurgery; in two cases radiofrequency surgery was used. All the children recovered without any complication (Katona et al., 2002). In 45 years tracheotomy was necessary in one infant only.

Kay and Goldsmith (2006) recommend a classification system for laryngomalacia which may be used as a surgical treatment strategy. Type 1 laryngomalacia is characterized by a foreshortened or tight aryepiglottic fold. Type 2 disease is defined by

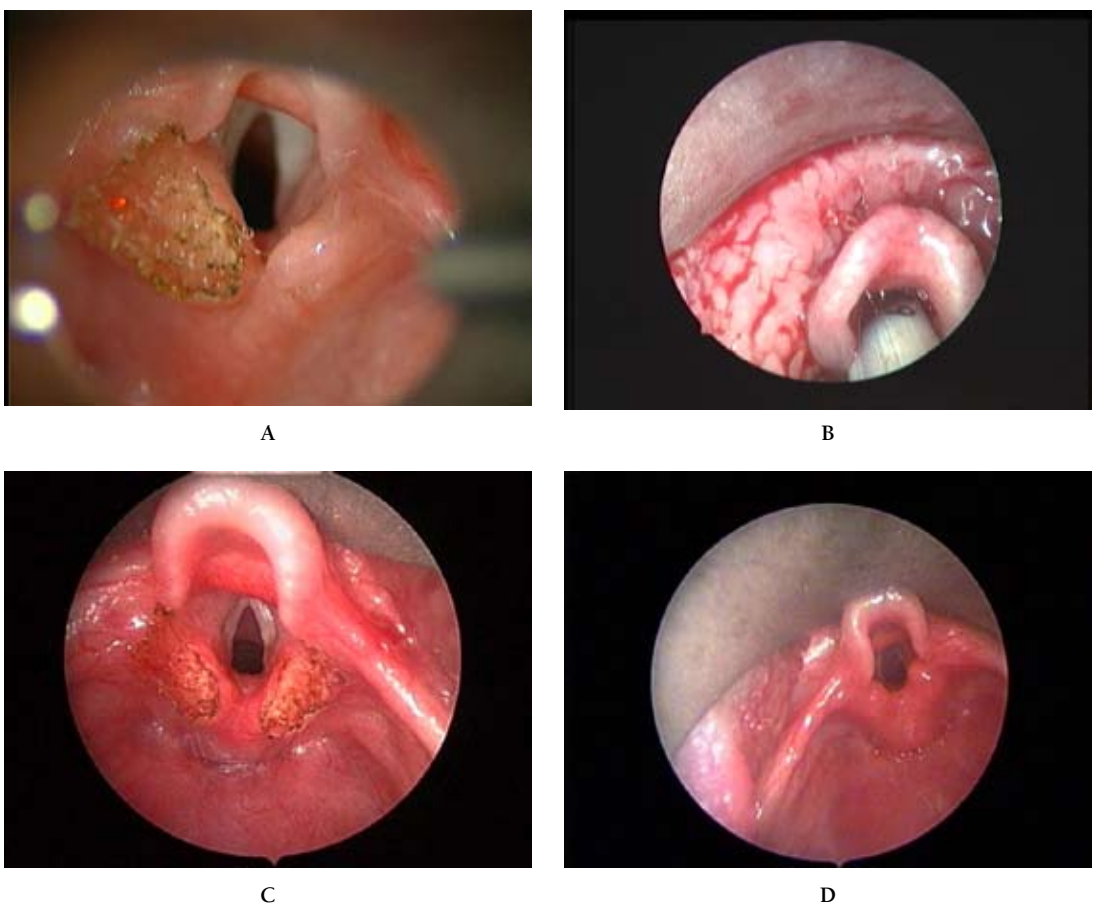


FIGURE 7-53. A. Laryngomalacia: laser supraglottoplasty (left side); **B**. epiglottopexy of epiglottis to the tongue base; **C**. supraglottoplasty—done; **D**. One month after supraglottoplasty.

the presence of redundant soft tissue in the supraglottis. The type 3 designation applies to cases caused by other etiologies, such as underlying neuromuscular disorders. Each type requires a specific approach to surgical repair. Stridor associated with poor feeding and failure to thrive may be the indication for surgery.

KEY POINT

Laryngomalacia is the most common cause of laryngeal stridor. The essence of this anomaly is congenital weakness of the laryngeal structure; thus, the designation: soft, flaccid larynx is more appropriate. Its main symptom is the cackling type of inspiratory stridor. Although this sound is very typical, laryngoscopy should be performed in all cases for verifying the diagnosis and to exclude other connatal anomalies with stridor. The crying sound is always clear. Laryngomalacia is a self-resolving condition (until the age of 1 to 1½ years), some kind of surgery is indicated rarely, only in severe cases.

To sum up: we can say that laryngomalacia is generally a self-resolving condition, but in severe forms endoscopic laryngeal surgery, according to some authors: should be performed, which is a safe method for treating this self-limiting condition (Merrot et al., 2004; Whymark et al., 2006). Among 30 children with severe laryngomalacia 83% obtained complete resolution and a further 7% an improvement after aryepiglottoplasty (Martin et al., 2005). If laryngomalacia is associated with additional congenital anomalies, failure of supraglottoplasty may more often be observed, in 8.8% among 136 cases, in the patient data of Denoyelle et al. (2003).

Atresia, Severe Stenosis of the Glottis

Laryngeal atresia is incompatible with life, unless recognized immediately after birth. Atresia means complete obstruction, yet the expression "partial atresia" is also used in the literature. It is meant to mean extreme stenosis at the level of the vocal cords, when the diameter of the glottis is a mere 1 or 2 millimetres (Baker & Savetsky, 1966; Ferguson, 1972a).

All kinds of sound production are absent in total atresia. When central nervous functions are preserved, the infant makes enormous efforts to breath: *unsuccessful respiratory motions, and a rapidly progressing cyanosis can be observed.* This condition is rarely diagnosed in vivo (Cohen, 1971; Kallay, 1974; Smith & Bains, 1966). According to Hicks, Contador, and Perlman (1990) total laryngeal atresia in newborns is a curiosity. We have seen only one case in 45 years. The (partial or total) atresia is often associated with other congenital anomalies, mainly with heart defects. The combination with 22q11.2 deletion (DiGeorge syndrome or velo-cardio-facial syndrome) was also observed (Fokstuen et al., 1997). *Only an urgent tracheotomy may promise minimal chance for survival.*

In excessive stenosis the vocal cords cannot be distinguished, and the lumen, if visible, is slitlike, not more than 1 to 2 mm in diameter. The infant is dyspnoeic, can hardly cry, producing only short, moaning sounds. *In partial but severe obstruction there is a phonation-like stridor, which may be noisy due to the unevenness of the surfaces causing the stenosis; the cry of these infants is wan, shrill, whistling, with pathologic changes in the acoustic structure* (Figure 7-54).

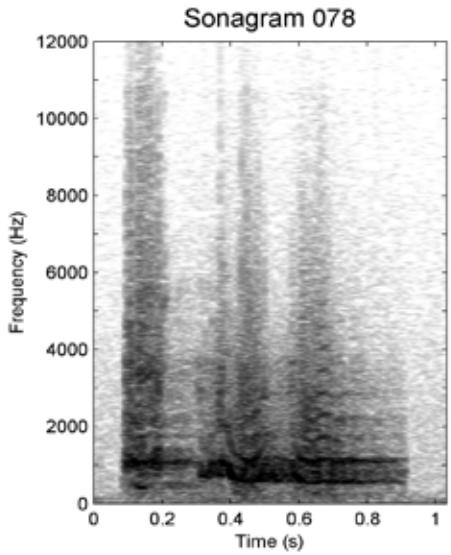


FIGURE 7-54. Severe congenital laryngeal stenosis. Infant aged 5 weeks. (For the *history* and current state of the patient see legend to Figure 6-14) *Acoustic finding:* Wan, whistling phonation (Fig. 6-14) and inspiratory stridor. *Sonagram* of the stridor *No. 78:*. Confluent noise throughout the entire spectrum; overtone-like components over 1500 Hz up to the highest frequencies making the stridor phonation-like. Between 500 and 1200 Hz there is a poorly divided noise component; the formant structures indicated above are also masked by noise. The inspiratory sound consists of two phases of roughly identical duration.

KEY POINT

Laryngeal atresia is incompatible with life. Unsuccessful respiratory motions, deep cyanosis may be observed, phonation is impossible. In partial but very severe obstruction wan, sometimes shrill crying sound and phonation-like, loud inspiratory stridor may be heard. Tracheotomy is unavoidable, later the transglottic severe stenosis can be operated on by laryngotracheal reconstruction.

In excessive severe stenosis the tracheotomy cannot be avoided. Before decannulation tracheoscopy is indicated and the eventually granulation around the internal opening of the tracheostoma should be removed (Figures 7-55 and 7-56).

In rare cases of survival reconstruction of the larynx may be advised..

Figures 7-57A through I show the surgical procedure of a severe (Grade 3) transglottic stenosis in a child aged 2 years: in the course of the laryngotracheal reconstruction laryngotomy and anterior and posterior cartilage graft were applied.

Prenatal diagnosis has been described; however, in many cases, the precise diagnosis is established only at autopsy (Minior et al., 2004).

Laryngeal Diaphragm (congenital laryngeal web)

This congenital anomaly of the larynx is most likely produced around the 10th week of intrauterine life by the same mechanism that causes laryngeal atresia and subglottic stenosis. Namely, the dissolution of the fused lateral masses

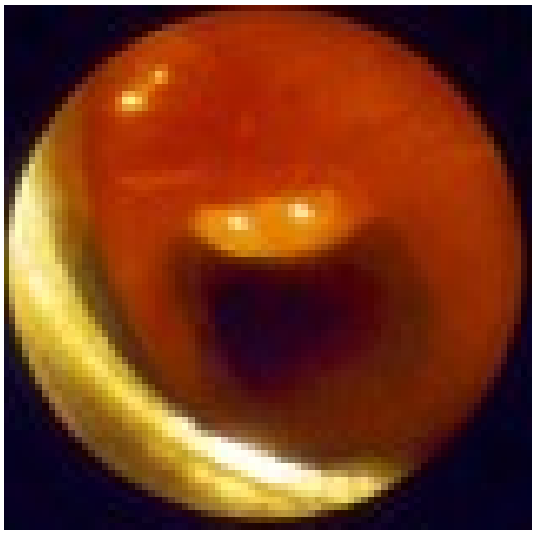


FIGURE 7-55. Granulation around the internal opening of the tracheostoma performed due to severe laryngeal stenosis.

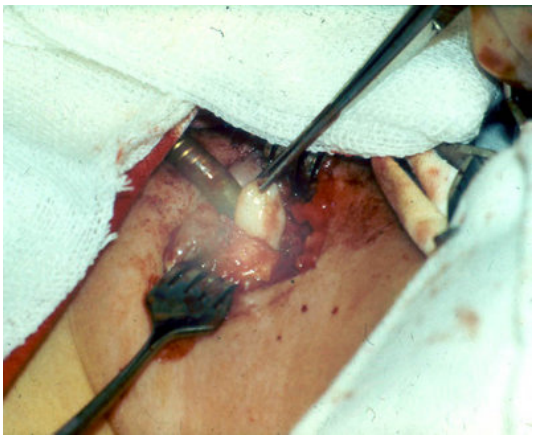


FIGURE 7-56. Removal of the tracheal granulation.

does not take place when the laryngeal lumen develops (Ferguson, 1972a).

The site of the laryngeal diaphragm (web) is mostly at the glottic level; in about 75% of cases (Holinger & Brown, 1967), less frequently above the vocal cords, and in about 20% it is subglottic in position. *The web may vary in thickness* (Lábas, 1968a) *from a membranelike, thin structure to a bulky bundle*. Its surface is covered by squamous epithelium, its posterior margin is typically concave. When it is situated between the vocal cords, the anterior two-thirds of the larynx are usually fused and occluded (Figure 7-58).

The symptoms and acoustic signs depend on the form, size, and location of the diaphragm. If the glottis is narrowed or the movements of the vocal cords are restricted, the cry is wan, shrill, and aphonic, like in congenital or neurogenic stenosis of the larynx, although it may also be clear and regular. *If the change is above the vocal cords or if it is subglottic, phonation is not affected*. Depending on the severity of the obstruction, respiration, too, may be impeded. *The character of the stridor depends on the location of the change*; in the case shown here stridor-phonation was observed. *If the web is subglottic, or a tapering web extends from the larynx toward the trachea, cough may also be present (nondescript cough or cough-phonation)* (Figure 7-59).

A third of children with laryngeal webs have associated anomalies of the respiratory tract, commonly subglottic stenosis, but an association between anterior glottic webs and velo-cardio-facial syndrome has been also observed (Tewfik & Sobol, 2003).

Diagnosis can only be established by rigid laryngoscopy and bronchoscopy; flexible endoscopy may reveal the presence of a laryngeal web but it is not always able to evaluate the extent of this anomaly. Unsuspected difficulties of intubation may call attention to the presence of a laryngeal web. Laryngography may be helpful

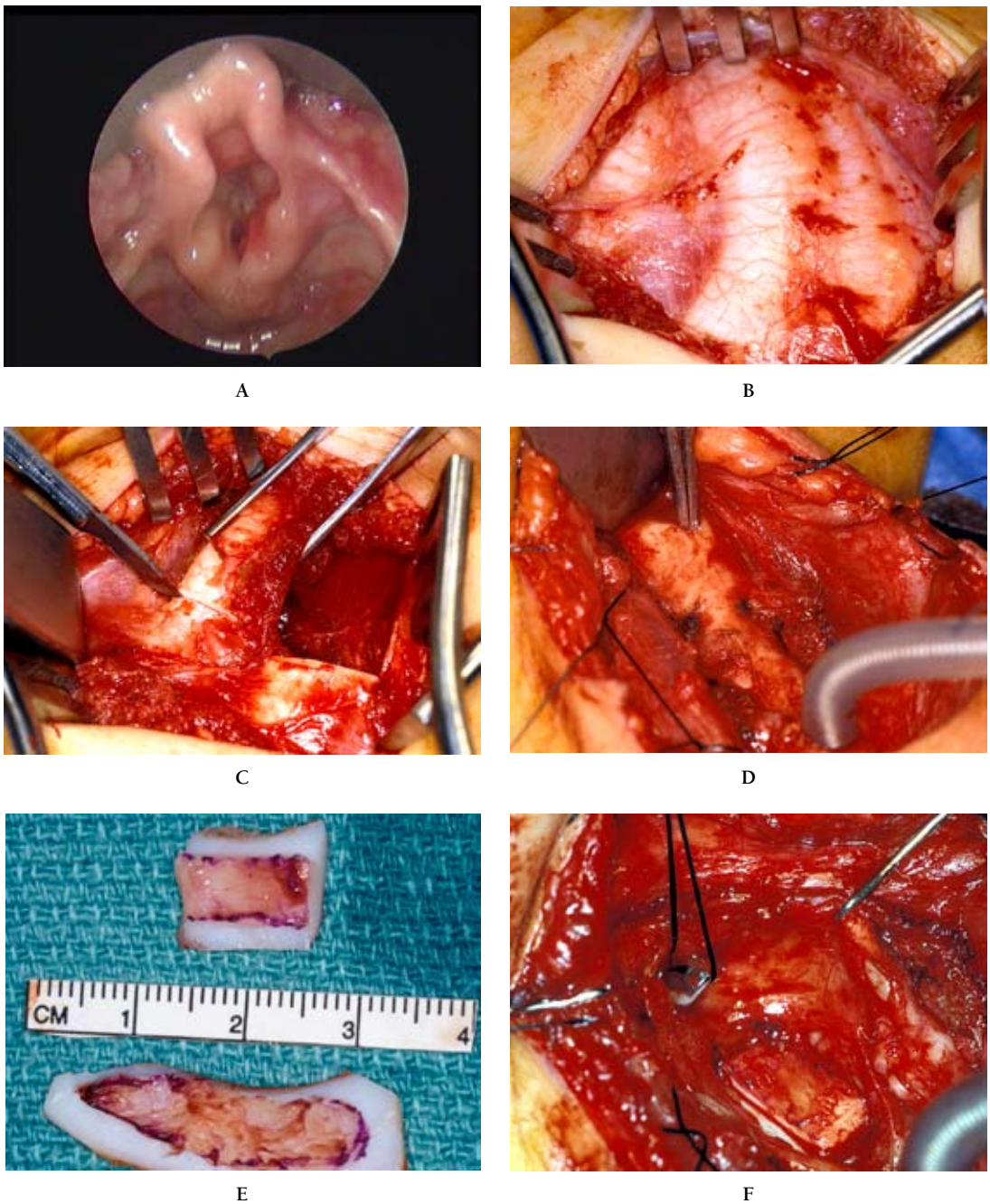
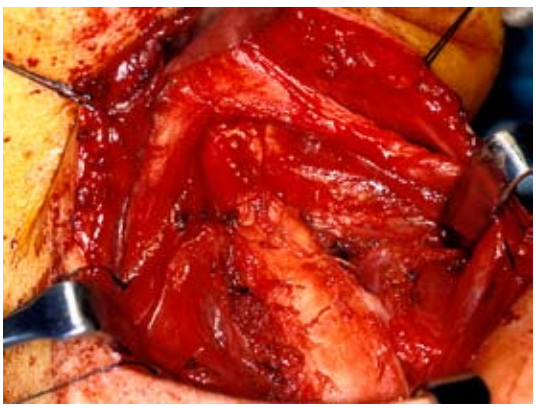


FIGURE 7-57. A. Transglottic stenosis high grade 3—infant aged 2 years. B and C. Laryngotracheal reconstruction. Anterior and posterior cartilage graft. D. Laryngotomy. E. Measure of grafts. F. Posterior cartilage graft, and G. anterior graft in place. H. 6 months postoperation. I. Transglottic stenosis: surgical drawing (Koltai).



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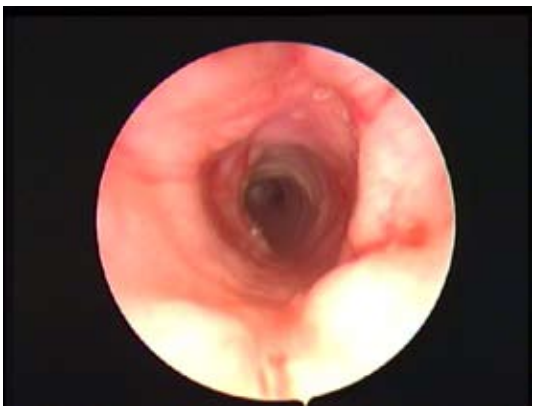

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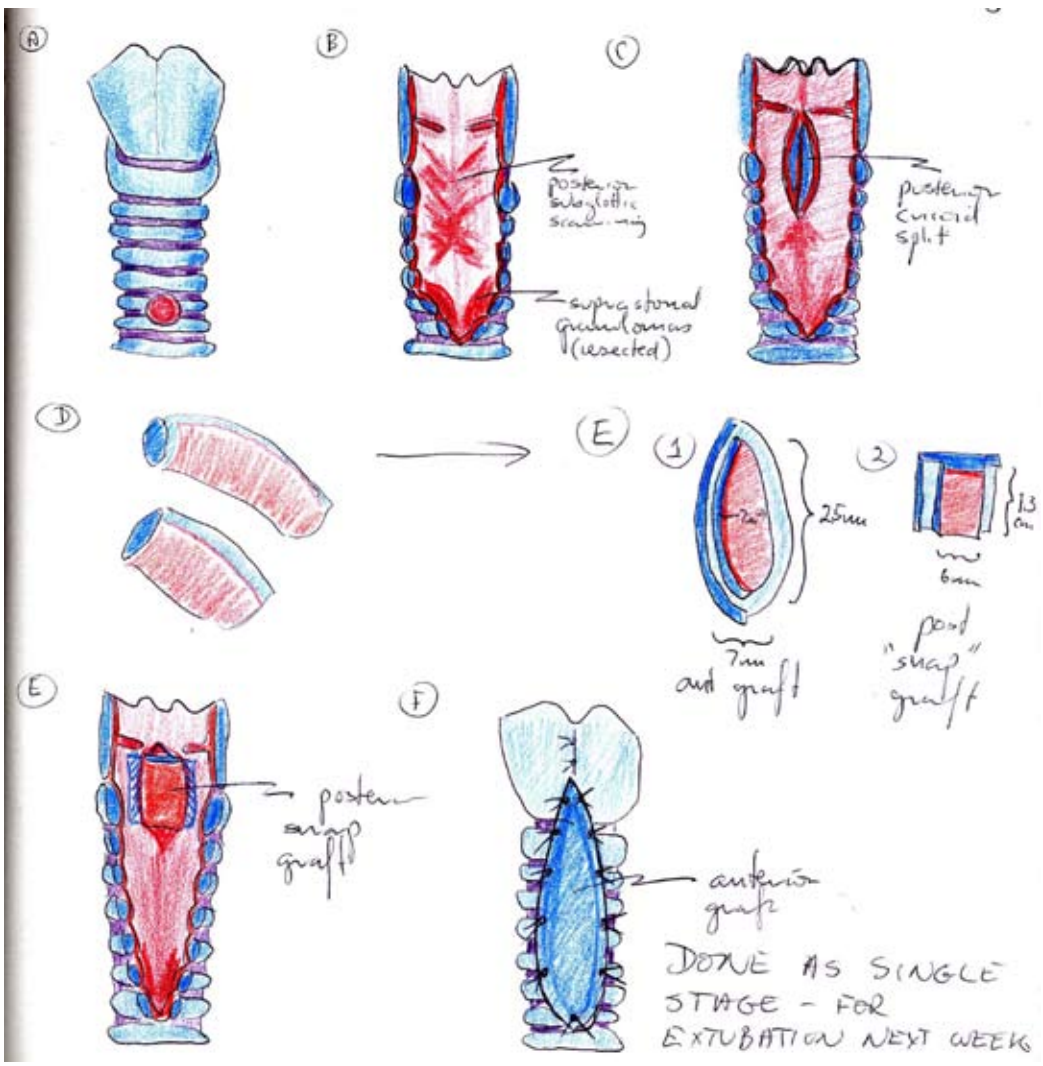

A
B
C
posterior subglottic scarring
supraglottal granulomas (resected)
posterior cricoid spl.t
D
E
1
2
25mm
13 cm
6mm
7mm
ant graft
post "swap" graft
E
F
posterior swap graft
anterior graft
DONE AS SINGLE STAGE - FOR EXTUBATION NEXT WEEK

i

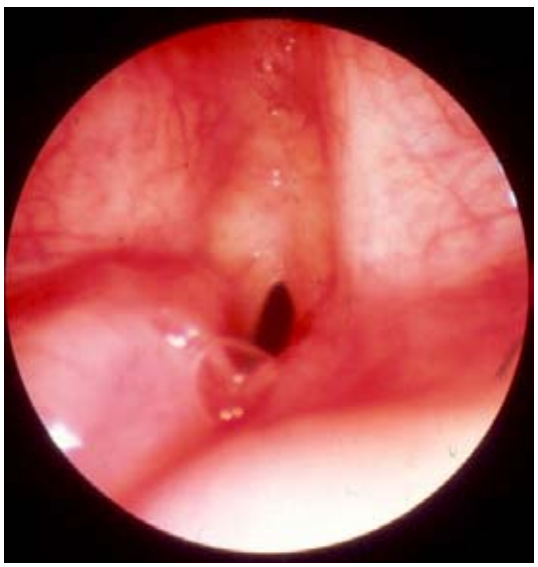


FIGURE 7-58. Laryngeal web in an infant aged 3 months.

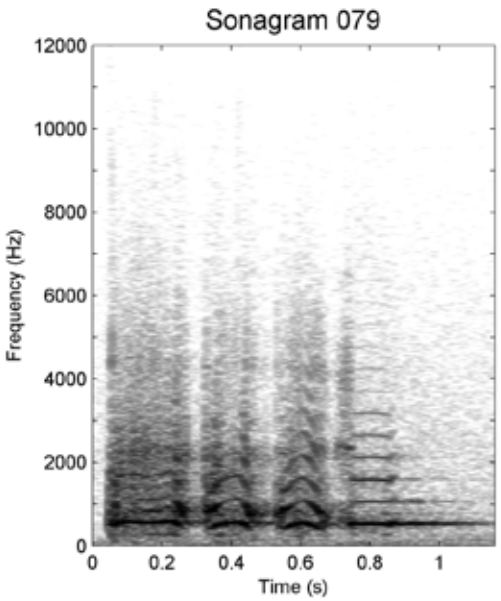


Figure 7-59. Laryngeal diaphragm (congenital laryngeal web). Child aged 20 months. *History:* Frequent episodes of dyspnea and cyanosis from birth. *Laryngoscopy:* There is a web between the vocal cords, occluding about half of the laryngeal lumen. *Laryngography:* The tapering web extends also into the subglottis. *Acoustic finding:* Stridor-phonation and nondescript, moaning cough. *Sonagram of the cough No. 79:* Confluent noise throughout the spectrum with the harmonic structure unfolding gradually during the three coughs. The overtones are roughly identical with the harmonics of the inspiratory stridor-phonation found at another part of the record. Slightly forced, moaning cough.

in determining the extent of the lesion (Figure 7-60). Radiographic evaluation is recommended to assess the site and measures of the laryngeal web prior to bronchoscopy. Lateral plain radiographs reveal a characteristic sail sign, representing persistent tissue between vocal cords and subglottis. High-kilovoltage technique is preferable because it gives better visualization of the soft tissue (Leung & Cho, 1999). Lateral neck radiograph should be taken during inspiration to avoid misdiagnoses. As laryngoscopy may fail to show the subglottic extension of the disease, Men, Ikiz, Topcu, Cakmacki, and Ecevit (2006) recommended recently *computed tomography and virtual endoscopy* which *may provide information needed also for surgery in such cases.*

Management of laryngeal webs ranges from observation to emergent tracheotomy. Thin, membranelike diaphragm may be operated on endoscopically using a cold knife or laser (Koltai & Mouzakes, 1999). Recently, Unal (2004) and Roh (2006) describe the successful management of congenital laryngeal web with endoscopic lysis and topical mitomycin C. Thicker webs require a laryngofíssure approach with postoperative stenting. Surgical correction usually entails expansion of the subglottic cartilage as well as lysis of the web across the membraneous vocal cord. Figures 7-61A through F show single stage laryngotracheal reconstruction with thyroid cartilage graft in a 1-year-old child with laryngeal web. Laryngeal web

FIGURE 7-60. Laryngography demonstrates the extent of the subglottic stenosis in an infant aged 13 months with tracheostomy.

excision with insertion of silastic keel also may be applied.

KEY POINT

Congenital laryngeal web may vary in thickness from a thin, membranelike structure (diaphragm) to a bulky, thick bundle spreading also into the subglottis and trachea. The symptoms depend on the size and location of the anomaly: the cry may be wan, shrill, or aphonic; inspiratory stridor and nondescript or barking cough may also be present. Diagnosis can be established with endoscopy, laryngography, x-ray methods, and MRI. The final management is surgery.

Vocal Cord Paralysis (abductor [recurrent] and adductor paresis)

The etiology of vocal cord paralysis may be divided into two categories of origin: congenital and acquired.

The *congenital paralysis* accounts for 10% of all congenital laryngeal lesions and it is the second or (according to other publications) the third most often occurring laryngeal anomaly (Friedman, de Jong, & Sulek, 2001), which cause symptoms immediately after birth.

Unilateral vocal cord immobility (*abductor or recurrent paresis*) is almost exclusively the result of peripheral nervous lesion (Ferguson, 1972b), because both halves of the larynx have central innervation. Connatal right-sided paresis is mostly due to overstretching or torsion of the neck during parturition. This is usually relieved spontaneously within a few weeks. The left-sided congenital paresis is mostly associated with cardiovascular malformations, for example, with ventricular and atrial septal defects or with enlargement of the heart in Ortner's syndrome. Unilateral recurrent (abductor) paresis may also be caused by a mediastinal process or cervical trauma (Kallay & Hirschberg, 1963). Pressure of neck terimes with various origin may also have a role in etiology.

The paralyzed vocal cords are mostly in a paramedian, less frequently in an intermediate position (Haguenauer, 1971) and they cannot be abducted on inspiration. The laryngeal lumen is narrowed to almost half its normal size, resulting in the production of inspiratory stridor-phonation. However, the paralysis may be incomplete, and the vocal cords may flutter slightly, when the stridor assumes a crowing character. *Mostly the cry is absolutely*

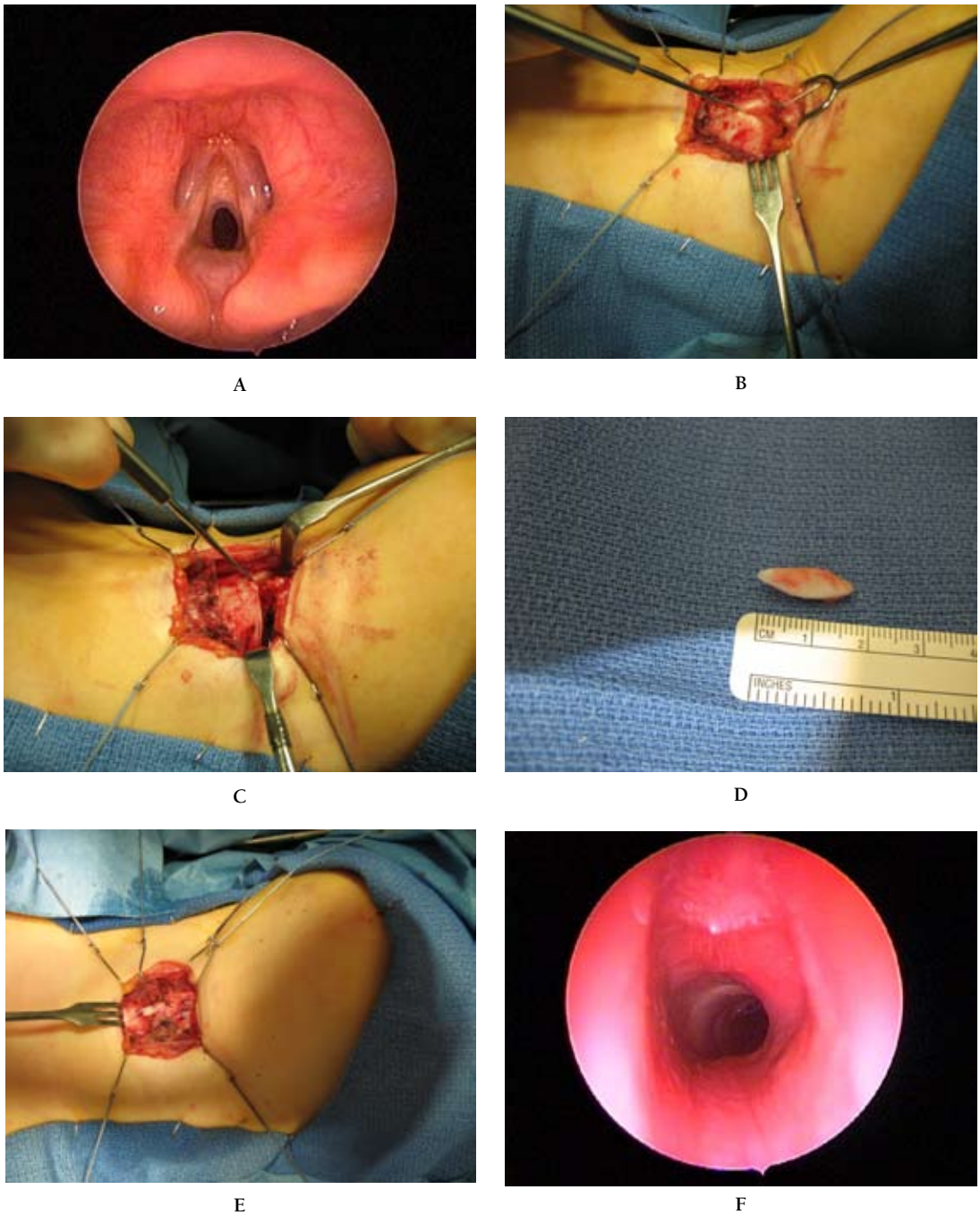


FIGURE 7-61. Congenital laryngeal web. Single stage laryngotracheal reconstruction with thyroid cartilage graft (A through E), (F) state 3 months postoperation.

clear and regular because, as a result of a compensatory mechanism, the contralateral vocal cord passes over the midline and bulging forward touches the paralyzed cord during phonation. Pathologic vocalization may also occur: *the cry may be shrill or creaking, or may tend to become sometimes weak, aphonic, or occasionally dull* (according to Haguenauer [1971], it is frequently bitonal), but we have never observed hoarseness (Figure 7-62). *Inspiratory or rarely biphasic stridor is present* that is louder when awake and improves when lying with the affected side down. According to Raes et al. (1982) except features of inspiratory stridor, no really clear parameters typical of the paresis of the recurrent nerve could be revealed in thirty infants. De Jong and Friedmann (2000) write in their publication that the most common manifestation of vocal cord paralysis is consistent stridor; ineffective cough, aspiration, recurrent pneumonia, and feeding difficulties can be associate
Bilateral paralysis or paresis is mainly of central nervous origin und much less uncommon than the unilateral case. It is the result of a lesion of the medulla oblongata, frequently associated with paresis of other nerves of the cerebral base. In the etiology intracranial hemorrhage, meningoencephalocele, hydrocephalus, neuromuscular diseases, and immature innervation may take place, but the cause of the paralysis cannot be cleared up in all cases. Unlike unilateral paralysis, the bilateral case produces severe respiratory impairment. The vocal cords are fixed in the paramedian position (Myer & Cotton, 1988; Réthi, 1955), almost completely ad-

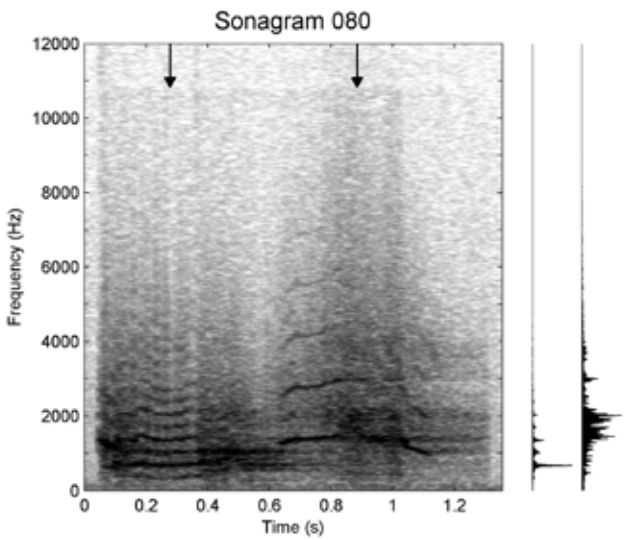


FIGURE 7-62. Right recurrent paresis. Infant aged 2 weeks. *History:* Stridor from birth. Disorders of swallowing and breathing (occasional "upper respiratory spastic dyspnea"). Transferred from maternity home. *Laryngoscopy:* Right vocal cord fixed in the paramedian position. *Acoustic finding;* Creaking cry, aphonia, stridor-phonation, and shrill cry. *Sonagram of the stridor-phonation and shrill cry No. 80:* 0.2 to 0.65 s: Inspiratory stridor. Normal overtone structure of about one-third of the inspiratory stridor (0.2 s), but only up to the 3500 Hz limit making the sound dull. Throughout the inspiratory phase a strong noise can be observed, particularly in the band between 500 and 1500 Hz; 0.65 to 1.1 s: Shrieking cry. Mutilated phonation composed of overtones at 1500, 2200, and 3300 Hz. The harmonic structure is masked by the strong noise of expiration. The shrieking cry is dull at the same time.

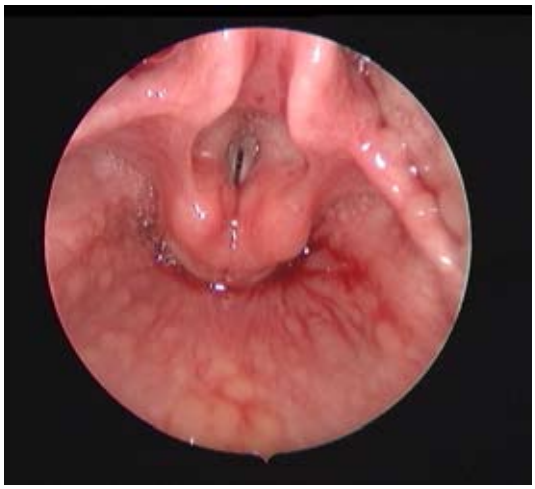


FIGURE 7-63. Bilateral vocal cord paralysis.

ducted, and the glottic slit, which may be oval in shape, is no more than 1 to 2 mm wide (Figure 7-63). Hardly any air can pass through this narrow slit.

Stridor in bilateral paresis is very marked even at rest (stridor-phonation of laryngeal origin is mostly completely free of noise or with a minimal superimposed noise; see Figures 7-64A and B). Phonation may be also severely impaired. *There may be almost total aphonia* or a defective cry tending to become aphonic. *In other instances the cry is shrill (with breaks and glides indicating the unstable flutters of the vocal cords), wan, or sizzling. Normal cry also may be observed*, because the paralyzed and weakly adducted vocal cords can vibrate during expiration and phonation, passively but steadily. Hoarseness (see Figure 6-6) is never found; the weak voice may occasionally even show a normal acoustic structure. Similar is the observation of Myer and Cotton (1988): the patient's cry may be normal but there is characteristically a high-pitched inspiratory stridor that worsens with stress.

The severe respiratory condition, the stridor of laryngeal origin and the weak, moaning vocalization or aphonia call for immediate laryngoscopy which will confirm the diagnosis. *The advent of the flexible fiberoptic nasopharyngolaryngoscope has brought a significant development in diagnostics*. However, it is not always easy to precisely determine the movements and position of the vocal cords in young infants, auditory observation and acoustic analysis may be of considerable help in making the diagnosis and planning the therapy. As a complementary method, ultrasound can be used (Vats, Worley, de Bruyn, Porter, Albert, & Bailey, 2004). In older children EMG may be useful for differential diagnosis. The latter may be especially important in acquired cases.

Acquired paralysis may have an infectious or traumatic origin. The vaccinations have greatly reduced the infectious type of vocal cord paralysis. Whooping cough encephalitis, diphtheria, tetanus, and botulism are rarely seen in today's pediatrics. Vocal cord paralysis due to endotracheal intubation is a controversial subject. A recurrent laryngeal nerve trauma can be suspected to pressure injury between the endotracheal tube and the superior portion of the thyroid cartilage. Thoracic and cardiovascular surgery may also cause injury of the recurrent laryngeal nerves.

Mechanical damage with severe restriction of the vocal cord movements may occur also in infants, possibly misleading the diagnostic procedure in some cases. In our case of a 2-month-old infant, no movement of the vocal cords could be seen for 3 months after a prolonged intubation. After 3 months a spontaneous recovery took place. That is the so- called

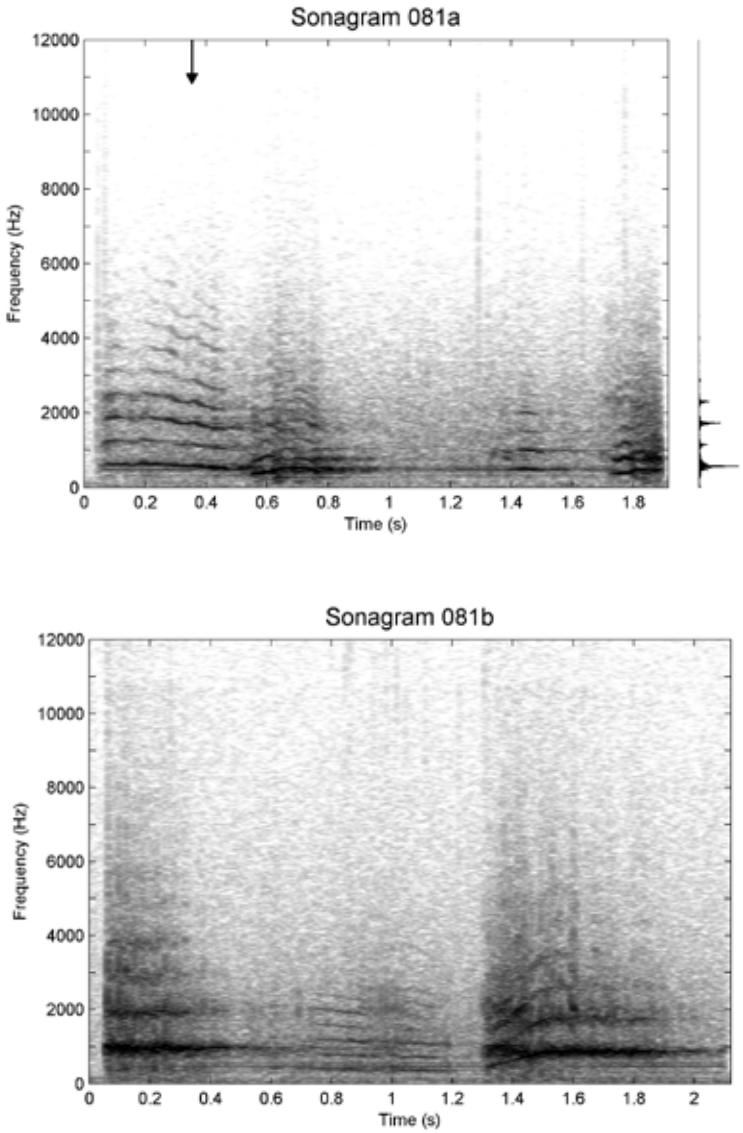


FIGURE 7-64. Bilateral laryngeal paresis. Infant aged 17 days. *History:* Cyanotic, severely dyspneic neonate weighing 4350 g, admitted with marked stridor in a moribund state at the age of 2 days. Cephalic haematoma, fracture of the clavicle, atelectasis of the lower lobe of the right lung. *Laryngoscopy:* Vocal cords fixed in the paramedian position, only the left one making slight movements. *Acoustic finding:* Faint, inert cry and whistling inspiratory stridor. **A**. *Sonagram No. 81a:* 0.3 to 0.7 s: Cry. Scanty harmonic structure disappearing at 3000 Hz. Weak fundamental frequency. Overtones around 1800 to 2000 and 2400 to 2800 Hz predominate. No substantial noise components. The intensity conditions of the overtones and fundamental (the higher overtones are more intensive than the lower ones and the fundamental) cause the inertia of the crying sound. **B**. *Sonogram No. 81b:* 1.2 to 2.0 s: Stridor-phonation in two respiratory cycles (starting with inspiration). Regularly arranged noise components up to 6200 Hz, with dominant bands between 600 and 900 Hz, 1500 and 2400 Hz, and at 5000 Hz. The relatively regular arrangement of noise components makes the stridor whistling and sibilant.

pseudolaryngeal paralysis (Figure 7-65). Severe posterior glottic stenosis could also be misdiagnosed as having bilateral vocal cord paralysis because of errors in observing the actual findings in the larynx: thus, the name pseudolaryngeal paralysis was given to these disease entities by Cohen (1981).

The essence of the therapy is the treatment of the basic disease; the task of the laryngologist is to ensure proper lumen of the airways. If the cause of the unilateral paralysis cannot be solved, and the later logopedic treatment is unsuccessful, the phonation may be improved with several augmentation techniques, for example, with collagen injection into the paralyzed vocal cord (Patel et al. 2003), but first in older children. Unlike the case in adults, careful identification of the vocal cord level should be performed in children before implant placement. Further research is necessary to prove prospectively that surgical intervention (e.g., thyroplasty techniques) in the pediatric larynx will not affect subsequent growth (Parikh, 2004). In bilateral paralysis tracheotomy is often necessary. In these severe cases (transitory or permanent) laterofixation of the paralyzed vocal cord may be performed (Lichtenberger, 1999, 2001; Lichtenberger & Toohill, 1997) (Figure 7-66). Arytenoidectomy with the CO_2 laser also has been used, but it has a failure rate of 20 to 40% because of postoperative scarring. Miyamoto, Parikh, Gellad, and Licameli (2005) treated 22 patients with bilateral congenital true vocal cord paralysis over a 16-year period at the Department of Otolaryngology and Communication Disorders, Children's Hospital in Boston; 15 of them required tracheostomy. Of the 15 tracheotomized patients, 10 were successfully decannulated (8 had spontaneous recovery, in two children a lateralization procedure was performed). Of the 7 patients not requiring tracheostomy, 6 recovered vocal cord function; 11 of the tracheotomized children had various comorbid factors, including neurologic abnormalities. Except for extreme infants (e.g., bilateral vocal cord paralysis with severe respiratory distress and central neuropathy) who require a temporary tracheotomy, Lee, Su, Lin, Tsai, Lin, and Lin (2004) recommend waiting at least 6 months before proceeding to invasive surgical interventions, as in their 13 patients younger than 1 year with vocal cord paralysis, spontaneous recovery occurred in 76.9% of the affected infants.

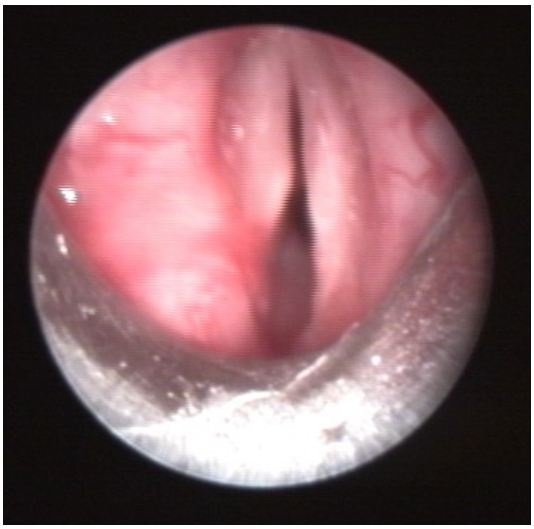


FIGURE 7-65. Pseudolaryngeal paralysis due prolonged intubation lasting 3 months in a child aged 6 months.

Case Study

A 5-year-old child with a very large cleft of the palate (see Figure 7-9), not operated on earlier, was sent to our depart-

ment for surgery. During the preoperative examination a severe inspiratory stridor could be observed when the child was moving. The uninformed parents believed it to be a consequence of the cleft palate. The speech of the boy was severily retarded and hypernasal due to the cleft. Laryngoscopy revealed bilateral vocal cord paralysis. No other neurologic alteration could be established (idiopathic case). After consulting several specialists various therapeutical possibilities came up: (1) observation only (but how long?), (2) first closure of the cleft in intubation narcosis: it was contraindicated by the anesthesiologists, (3) first develop and improve the speech, breathing can be a secundary problem (viewpoint of some speech therapists), (4) tracheostomy (how and when can the child be decannulated?), (5) botulin toxin injection (pediatric neurologists were strongly opposed, all the more because this management has to be repeated every 4 to 6 months), and (6) the treatment sequence finally accepted by us was as follows: tracheotomy, closure of the palate, and then laterofixation of one vocal cord (see Figure 7-66), decannulation. After 1 year the child had no respiratory problem, his voice was normal without any hoarseness (due to great compensation abilities in childhood), and his speech rapidly developed without hypernasality.

Bilateral *adductor vocal cord paralysis*, presenting itself with features of laryngeal incompetence, is very rare in infants (Berkowitz, 2003; Ferguson, 1972b); in three cases of Berkowitz (2003) it was associated with Robin sequence and 22q deletion. We have not found this in any of our cases.

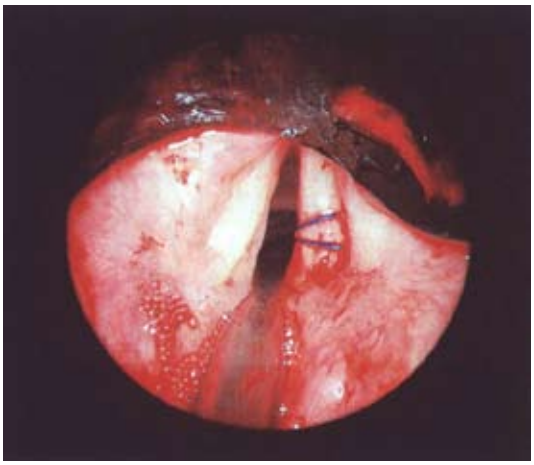


FIGURE 7-66. Laterofixation of the right vocal cord sec. Lichtenberger in a child aged 4 years with bilateral congenital vocal cord paralysis.

KEY POINT

Vocal cord paralysis may be connatal or acquired, unilateral or bilateral. When unilateral the cry is shrill or creaking, but absolutely clear cry may also be observed; besides, a typical laryngeal inspiratory stridor occurs.When bilateral, the stridor is very marked also at rest; the crying sound is extremely vacillating, from aphonia to normal, regular cry, depending on the anatomic situation. Diagnosis can be made only by endoscopy, but ultrasound, electromyography, auditory, and acoustic evaluation may help in diagnostics. As for the various forms of paralysis and the therapy see the text.

Dysphonia

Dysphonia; that is, forced, pressed, spastic sound production, may occur transiently in healthy, normal infants, as has been pointed out in several reports (Lind, 1965; Ostwald, 1972; Sedláčková, 1967a). Pathologic dysphonia, on the other hand, is either congenital or persistent. It is a common disorder in adults (Frint, 1982; Perelló, 1962), and children. According

to Kittel (1984) the disturbance of vocal purity occurs at least at certain levels in 23% of all children: dysphonia is the most important laryngeal symptom. *Dysphonia is,* however, *not identical with hoarseness*. Dysphonia is a superordinate category, a cover term ("Oberbegriff"): *hoarseness may be its leading symptom, but it is characterized also by the change of tone-color, melody, and resonance* (Frint & Hirschberg, 1979; Kittel, 1979; Wendler, Seidner, & Eysholdt, 2004).

KEY POINT

Hoarseness and dysphonia are not synonyms. Dysphonia is a superordinate category, it is characterized, besides hoarseness, also by structural changes of the voice spectrum: sudden changes in pitch or melody, glides, breaks, bitonality, or aphonia may occur.

On the other hand, *infantile dysphonia is uncommon* and its various forms have been first described by one of the present authors (Hirschberg, 1970, 1985). Parents of such infants first seek medical advice because of the strange voice production they observe. Crowe and Zeskind (1992) investigated perceptual and psychophysiological responses of 30 nonparent adults to various infant cries. They concluded that the observers found the hyperphonated (dysphonic) cries to be more aversive, distressing, urgent, arousing, and sick than phonated (normal) cry sounds. *The cry in dysphonia may be highly variable, but stridor cannot be observed in any of the cases.* As laryngoscopy mostly fails to reveal any relevant alteration, auditory observation is of particular importance in these cases, just like the acoustic analysis of the cry. The auditory experience may be a "hoarse" sound, but the disorder described by us as "hoarse cry" has never been demonstrated; it is not noise, but the structural changes of the voice that are relevant.

KEY POINT

Dysphonia in newborns is uncommon, it is a consequence of the immature innervation of the vocal cords mainly due to latent injury during birth or encephalopathia. The dysphonic cry is many folded: in hyperfunctional form pressed, tense, raucous phonation may be heard, whereas in hypofunctional form faint, inert, wan cry is typical. The two basic forms can occur in combination with each other. Stridor is never present: this observation is important as for differential diagnosis (e.g., to distinguish from laryngeal paralysis). Laryngoscopy cannot reveal any alteration. Infantile dysphonia can be over in some weeks spontaneously.

We have pointed out in our earlier reports (Hirschberg, 1970, 1973b, 1974) that *dysphonia may be either hyperfunctional or hypofunctional. In the background of both there may be a latent injury at birth, mostly encephalopathy, which causes immature innervation of the vocal cords* (Hirschberg, 1999). In its hyperkinetic type the change in phonation is secondary resulting from the agitation and aggressivity associated with brain damage, that is, from much crying and shouting. Hypokinetic dysphonia may be a direct consequence of the encephalopathic involvement of the corticonuclear tracts participating in the nervous control of sound production. One might also suggest that the symptoms of hypofunction are produced by a bilateral lesion of the superior laryngeal

nerve causing paralysis of the laryngeal tensor muscles.

The acoustic attributes of the two types of dysphonia are not always unequivocal. *In the hyperfunctional type there is usually a pressed, tense, raucous or crackling cry, whereas the faint, inert, or wan cry is subjectively found in the hypofunctional type*. Hyperfunctional state of the supraglottic area is indicated by pressing whereas that of the vocal cords by a tense cry. If there is a concurrent increase in subglottic pressure, the fundamental leaps an octave and a shrill sound is produced. A shrill, whistling, sharp voice also may occur as a compensation for a faint, wan cry; when the shrill sound is high-pitched at the same time, the cry becomes very thin or creaking, with irregular frequency variations.

The dysphonic voice may be characterized by sudden changes in pitch, glides, melody changes, sudden breaks, bitonality, or aphonia. At the same time, also clear, regular cry may occur, indicating that the pathologic change is not local but a disorder of central innervation.

The highly variable cries found in dysphonia are illustrated by Figures 7-67, and 7-68. When the condition improves and the neurologic symptoms subside, phonation, too, spontaneously becomes normal in some weeks (Hirschberg, 1990a, 1990b, 1998a, 1998b), as can be established both auditorily and by acoustic analysis (Figures 7-69A, B, and C).

It is a very rare exception that the infant dysphonia extends over childhood; *dysphonia in children is more frequently an independent clinical entity in impulsive children,* due to vocal abuse or disuse. In the case of a long-lasting dysphonic voice production *a vocal cord nodule may develop* (Figure 7-70).

Shah, Woodnorth, Glynn, and Nuss (2005) evaluated 646 patients with videostroboscopic examination and perceptual analysis of voice characteristics from 1996 to 2003, who were sent to the tertiary care pediatric hospital's voice center; 254 patients (40%) with an average age of 7.7 years were identified as having vocal nodules; 6 patients were under the age of 7 months. According to the authors evidence of gastroesophageal reflux disease was found in one-quarter of the patients; hyperfunction of the larynx was seen in three-fourths. The severity of vocal changes correlated with the size of vocal nodules.

There is no unanimous standpoint regarding the surgical removal of the vocal nodules in childhood (Hirschberg et al., 1995), but in our practice the excision may be occasionally necessary and helpful in older children (Hirschberg, 1996; Hirschberg & Szabó, 1965). Surgical therapy was performed by us in 6% of our 1890 dysphonic children aged 5 to 6 years on average during 10 years (Hirschberg, 1985, 1998a; Hirschberg et al., 1995). According to our experience surgery may be indicated (1) in cases with vocal cord cyst or polyp, (2) when speech therapy fails in at the latest, 4 to 6 months, or (3) in patients with very large, hard nodules if clear voice can not be developed even with special techniques (Figures 7-71A, B, and C).

Hyperbilirubinemia

Bilirubin plays a major role in the development of central nervous system pathology (Vohr et al., 1990). Transitory hyperbili-

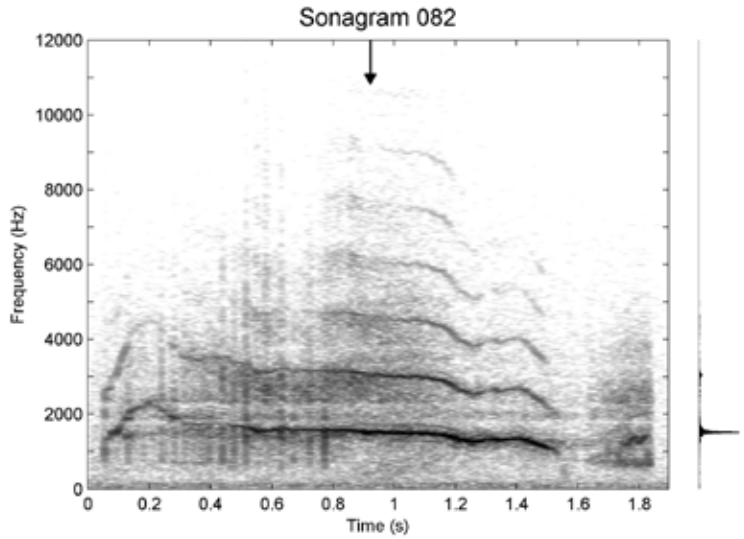


FIGURE 7-67. Dysphonia. Infant aged 17 months. *History:* Following plastic surgery of the lip under intubation anaesthesia the infant was found to have hardly any voice. Intubation complication suspected, tracheotomy contemplated. On grounds of the acoustic pattern, the absence of voice was believed to be central in origin, as confirmed by the additional data of history referring to a weak cry ("thin voice") from birth. *Acoustic finding:* Creaking, very thin crying sound, no stridor. *Sonagram No. 82:* No component corresponding to the normal fundamental frequency can be observed; instead, a relatively high overtones appear, of which even the lowest is 1700 Hz, or occasionally 2500 Hz. Scanty, mutilated harmonic structure: at some parts there are only two overtones. Masking by noise varies, but is not marked. The scattered overtones occasionally show considerable variations in melody (0.1–0.4 s); between 2500 and 5000 Hz the second overtone glides 2500 Hz within 0.15 s. The sound is high-pitched at the same time shrill. It is also very thin and creaking.

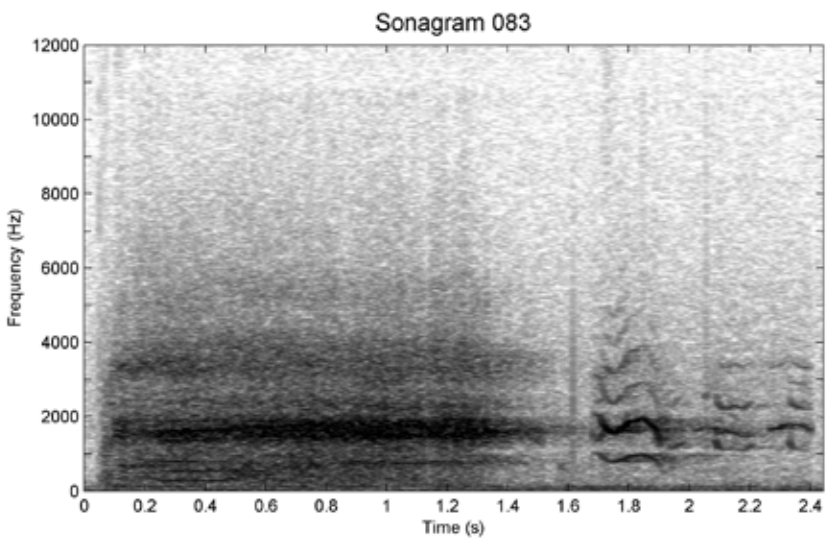


Figure 7-68. Dysphonia, false-vocal-cord sound. Infant aged 4 months. *History:* "Rattling" in the throat and difficult breathing from birth. *Laryngoscopy:* No pathology. *Acoustic finding:* Pressing sound production. *Sonagram No. 83:* Regular noise bands around 1500, 2300, 3500, and 5300 Hz produced by abnormal false-vocal-cord activity. No phonation develops beside these intensive components. (Dysphonia is followed by three brief phonations of regular acoustic structure.)

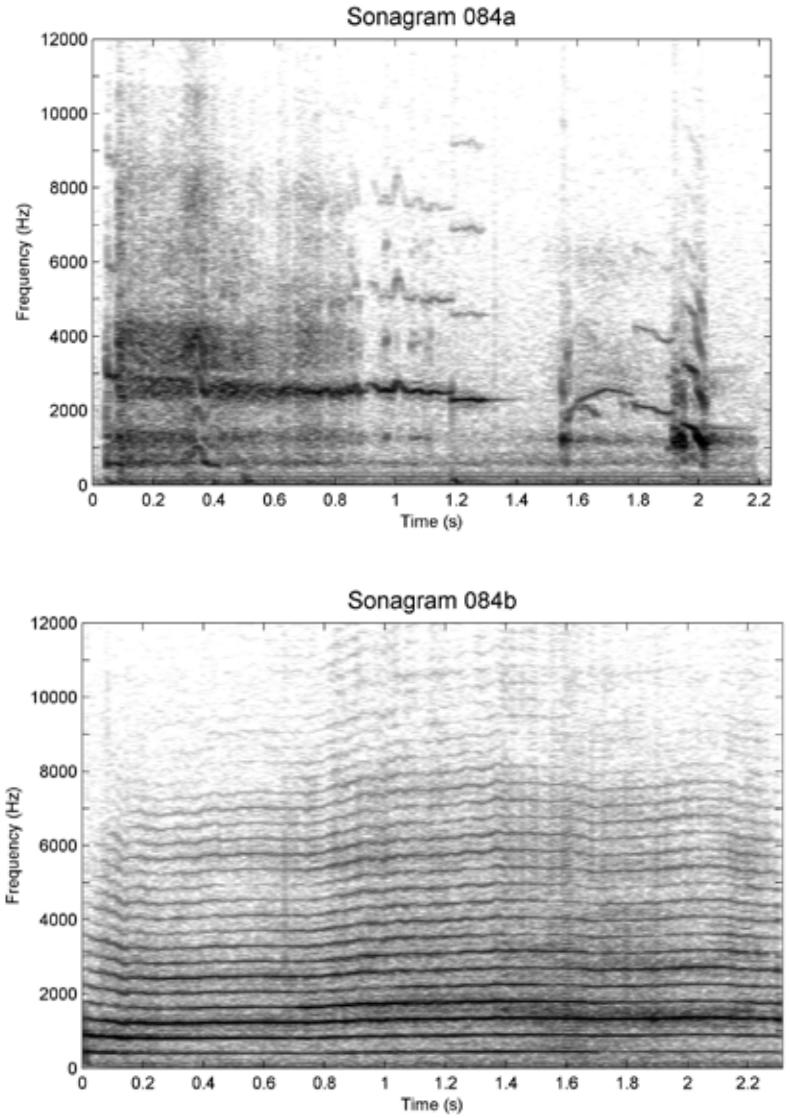


FIGURE 7-69. Encephalopathic dysphonia. Infant aged 5 days. *History:* Reported to have been "crying silently" ever since birth. Turns blue sometimes during feeding. Reduced turgor, acrocyanosis, parchmentlike, grayish skin. *Laryngoscopy:* Without any pathologic change. *Acoustic finding:* Shrieking, breaking, occasionally aphonic voice (A). Sound production normalized presumably with improvement of the cerebral symptoms (record B made 5 weeks later). **A**. *Sonagram No. 84a:* Amorphous, noisy, and hoarse sound which fails to create a phonation impression; at 0.9 s three overtones emerge showing a 400 Hz glide within 0.05 s at, for example, 1.05 s and an abrupt break of about 350 Hz at 1.2 s. After a segment of aphonia (1.4–1.55 s) the same structure recurs but with lower intensity (until 2.0 s). The three overtones appear at 2500 Hz, between 4500 and 5500 Hz, and between 7500 and 8000 Hz. In the shrill part of the cry there is no noise, but the overtones are markedly thickened. Shrieking, piping, occasionally breaking cry of long duration, turning into aphonia at sites. **B**. *Sonagram No. 84b:* Regular harmonic structure with the overtones about 480 Hz apart. Low-intensity noise component between 5000 and 6500 Hz. Normal cry. **C**. *Minimal time interval spectrum* of the abnormal cry. Irregular noise structure, with variable arrangement of the components.

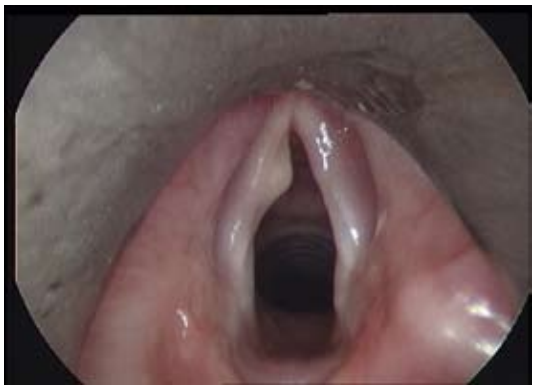
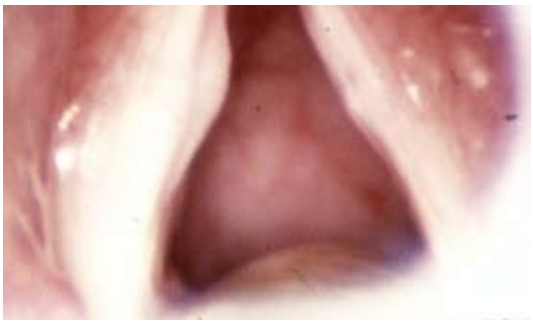


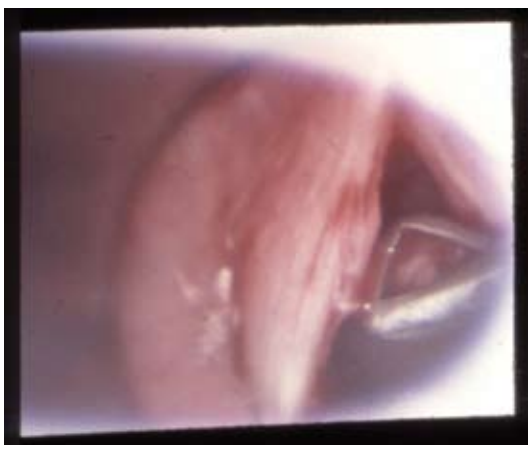
FIGURE 7-70. Vocal cord nodule in a 6-year-old child.

rubinemia is not a rare condition in neonates. It increases in severity in the first 35 days, decreasing thereafter and returning to the normal value during the second week of life. In more than half of the cases bilirubinaemia is slight, less than 34 µmol/L, and jaundice is not observable on inspection (Petényi, 1961). Physiologic jaundice refers to jaundice in the immediate newborn period without signs of illness (Parks, Montgomery, & Yetman, 2006). In cases of Rh incompatibility and isoimmunization, the serum bilirubin level may be as high as 340 to 850 µmol/L, and then kernicterus also may occur. Jaundice is intensive on the first or second day in these cases, the infants' general condition deteriorates, it becomes apathic, and sensory disorders develop. At present, the up-to-date management of the newborns prevent these severe symptoms in most cases, but it has not disappeared entirely. The significant decrease of the serious sequelae is undoubtedly the result of a successful prevention.

Newman et al. (1999) reported that 2% of the newborns had total serum bilirubin levels higher than 20 mg/dL, 0.15%



A



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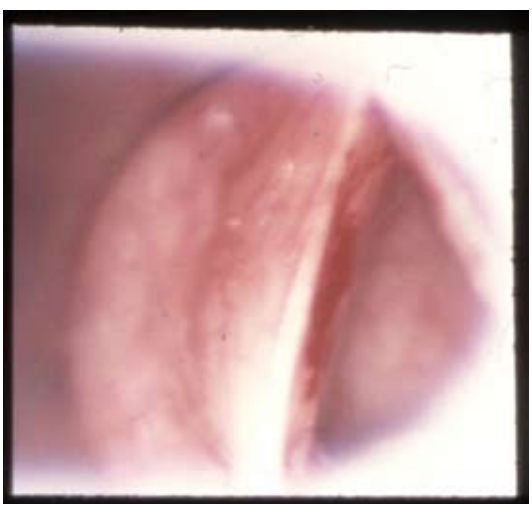


C

FIGURE 7-71. A. 4-year-old child with dysphonic nodule on the left vocal cord. **B.** During excision of the vocal nodule. **C.** After surgery.

had levels higher than 25 mg/dL, and only 0.01% had levels higher than 30 mg/dL among 50,000 California infants examined between 1995 and 1996.

The American Academy of Pediatrics requested recently an evidence-based report from the Agency for Healthcare and Quality that would critically examine the available evidence about hyperbilirubinemia and kernicterus (Ip et al. and Subcommittee on Hyperbilirubinaemia, 2004). From the evidence-based review it was evident that the preponderance of kernicterus cases occurred in infants with a bilirubin level higher than 20 mg/dL. Kernicterus is a pathologist's term describing the yellow desposits and brain cell death from extremely high levels of bilirubin, generally >2530 mg/dL. The term is often used interchangeably with bilirubin encephalopathy. Hyperbilirubinemia, in most cases, is a necessary but not sufficient condition to explain kernicterus. Factors acting in concert with bilirubin must be studied to seek a satisfactory explanation. Kernicterus, although infrequent, has also today at least 10% mortality and at least 70% long-term morbidity (Ip et al., 2004).

Hyperbilirubinaemic neonates may produce a very high pitched, shrill cry (Figure 7-72). In earlier literature on kernicterus it was mentioned that an even more specific acoustic phenomenon, so-called furcation, may occur (Wasz-Höckert et al., 1971). This means a peculiar break in phonation: the high fundamental appears as a series of harmonics. Furcation has been observed at the onset or end of the cry, and is derived from an abnormal vocal cord activity. On the basis of the examina-

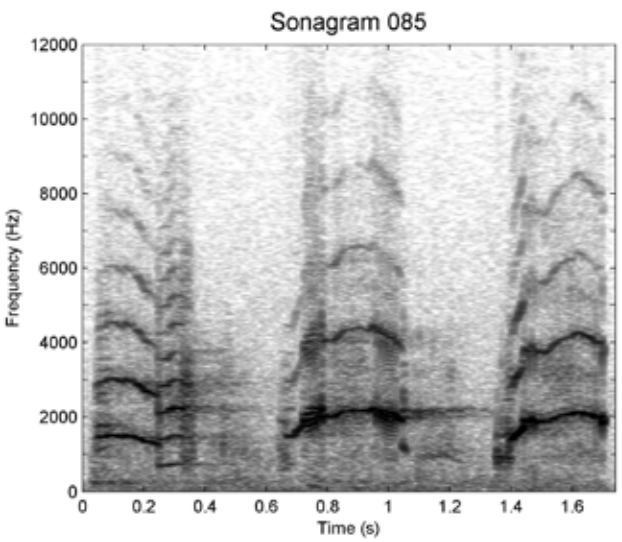


FIGURE 7-72. Hyperbilirubinemia (serum bilirubin: 268 µmol/L). Small-for-date newborn (2150 g) aged 6 days. *Acoustic finding:* High-pitched, shrill cry. *Sonagram No. 85:* Scanty harmonic structure up to the highest analyzed frequency components. The lowest frequency values are seen in the frequency domain of 750 Hz. Dominant overtones show up around 1500 and 2000 Hz. and the rest at 3000, 4000 to 4500, and 6000 to 6500 Hz. Considerable masking by noise and thickening of the individual overtones. The whole sound picture is in three ǟ = 0.3 s wide blocks apart having modulations in the 1000 to 1500 frequency Hz range, and especially in the 15th and 16th

tion of 50 term infants Vohr et al. (1989) concluded that hyperbilirubinemia affects the cry production: the moderate serum bilirubin group (10 to 20 mg/dL, 170 to 342 µmol/L) results in an increase in percent of cry phonation and an increase in the variability of the first formant in comparison with the low serum bilirubin group.

Treating hyperbilirubinaemia will likely prevent kernicterus in most instances; total serum bilirubin (TSB), however, is a poor indicator of kernicterus risk; thus, we need to explore the use of newer tools in conjunction with TSB to target infants at higher risk (Ip et al., 2004).

KEY POINT

Transitional hyperbilirubinemia in neonates is not a rare condition but in cases when the serum bilirubin level is high and the consecutive jaundice will be prolonged, the infants' general condition worsens. They become apathic and also a very high-pitched, shrill cry may be observed. This condition is no more common today due to efficient prevention. Once, pediatricians could establish the diagnosis of brain damage on the basis of this peculiar crying sound.

In the prevention of severe consequences of hyperbilirubinemia blood exchange transfusion and phototherapy may come up. Phototherapy has an absolute risk-reduction rate of 10 to 17% for prevention of serum bilirubin levels higher than 20 mg/dL in healthy infants with jaundice. Based on the results from the most recent study to report blood exchange transfusion (BET) morbidity, the overall risk of permanent sequelae in 25 sick infants who survived BET was from 5 to 10%. According to Jackson (1997), because of the significantly higher rate of severe complications in ill infants, exchange transfusion should be delayed until the risk of bilirubin encephalopathy is as high as the risks of severe complications from the procedure itself (12%). Newer phototherapy techniques are likely to reduce the need for BET (Maisels, 2001). Phototherapy combined with cessation of breastfeeding was found to be the most efficient treatment protocol for healthy term or near-term infants with jaundice (Ip et al., 2004; Martinez et al., 1993).

Down Syndrome (mongolism)

Down syndrome is the most frequently occurring chromosomal abnormality. It was the first well-defined syndrome from among a heterogeneous group of mental retardations which Langdon Down described in 1866 (Czeizel et al., 1973).

KEY POINT

Down syndrome is the most common chromosomal abnormality. The symptoms (the clownlike outward appearance and muscular hypotony) are sufficiently significant for making a diagnosis which can be confirmed by chromosomal analysis. The phonation is also very typical: the cry is prolonged, bleating, pressed, and low-pitched. The usually enlarged tongue may result in pharyngeal stridor. Acoustic procedures obviously demonstrate these changes in newborns.

The most common symptoms include excessive muscular hypotony, hyperflexibility of the joints, loss of the Moro reflex, a flat profile, oblique eyeslit, short, broad neck, malformation of the hands, dryness of the skin, and mental retardation. The

facial traits and the peculiar, clownlike expression are typical; intelligent gaze and fixation are absent even after 6 months of age, and grimacing often may be observed. Most children with Down syndrome have IQs that fall in the mild or moderate range of retardation. Some are so mildly affected that they may live independently and are gainfully employed.

The symptoms are sufficiently characteristic to make the diagnosis which can be confirmed by chromosomal analysis (trisomy of chromosome 21).

The phonation of the infants with Down syndrome is so characteristic that the diagnosis can be established with reasonable certainty by acoustic analysis, even before the results of the lengthy chromosomal analysis become available. The cry is bleating and pressed, creating the impression of being low-pitched. The abnormal prolongation of the cry, as well as interruptions of longer or shorter duration occurring in the same phase of the cry are typical features (Figure 7-73). Lind et al. (1970) observed in the cry a characteristic feature, called "stuttering" which is produced by rapid, rhythmic variations in glottal pressure. The mechanism of the pathologic cry is still unclear (Lind et al., 1970).

Pryce (1994) performed an interesting investigation using an electromyographic biofeedback technique to measure the tension in the muscles surrounding the larynx in adult patients with Down syndrome. The purpose of her study was to explore possible causes of the often acknowledged harshness, hoarseness, and gruffness in the voices of people with Down syndrome (Wilson, 1987). The hy-

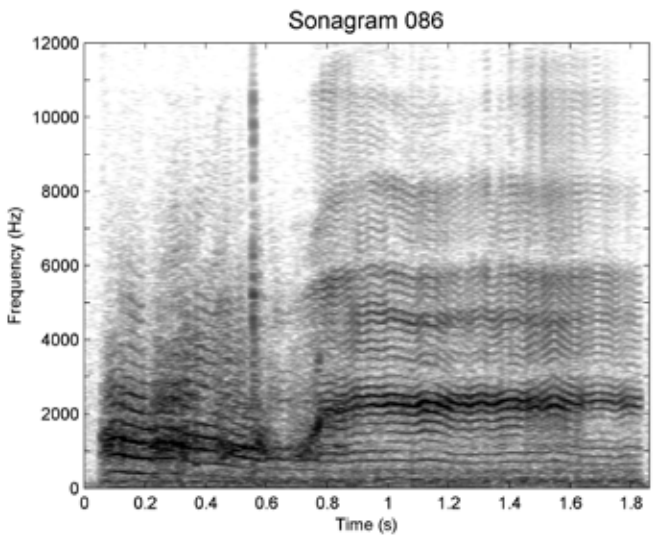


FIGURE 7-73. Down syndrome. Infant aged 3 weeks. (For the *history* see legend to Figure 6-17*) Acoustic finding:* Prolonged (9 s), pressed, deep, bleating cry. *Sonagram No. 86.* (showing the first part of the prolonged cry): The characteristic acoustic structure of normal cry first (regular harmonic structure, although the noise components characterizing the pathologic cry tend to appear also in the initial stage), which gradually disintegrates and is replaced by a regular noise structure. Two intensive, broad bands of noise appear around 2500 and 5800 Hz. At the same time, a fundamental vibration giving the cry its bleating character arises (see also Figure 6-17). The sound

pothesis that hypotonia (reduced muscle tone) gives rise to a floppy laryngeal musculature in this anomaly, was not proven; the chief finding was that the energy level needed to activate the vocal mechanism from its at-rest level to its voicing level is almost twice as great for the group with Down syndrome as for the control group.

Due to the usually enlarged tongue, the pathologic cry may be accompanied by pharyngeal stridor.

Cri du Chat Syndrome (cat's cry disease)

The chromosomal abnormality underlying this congenital anomaly is the deletion of the short arm of the fifth chromosome (Lejeune et al., 1963). The principal symptoms are craniofacial dysmorphism with microcephaly, somatic and mental retardation, feeding problems, behavior problems such as hyperactivity, aggression, and a crying sound similar to the mewing of the cat. The latter typical symptom has given the disease its name.

The pathologically altered cry has been extensively studied (Altrogge et al., 1971; Bauer, 1968; Goydke & Schroeder, 1972; Luchsinger et al., 1967; Schroeder et al., 1967; Sohner & Mitchell, 1991; Šubrt et al., 1969; Vuorenkoski et al., 1966).

The cry is high pitched, with a scanty harmonic structure and undulating overtones making it sound shrill and mewlike. There is a striking similarity between the spectrographic pattern of the cry of cri du chat syndrome infants and the mewing of young cats (Figures 7-74A and B). The crying phrase is usually long and less periodic than normal; the melody form is often flat, making the cry monotonous. Sohner and Mitchell (1991) observed similar acoustic signs: high fundamental frequency and predominance of falling intonation.

There are two hypotheses to explain the production of this pathologic cry: according to some authors (Fazekas & Szemere, 1972; Truffer, 1970; Woldorf & Pastore, 1972) a local change, that is, some abnormality of the laryngeal cartilages, would be responsible, whereas others (Altrogge et al., 1971; Schroeder et al. 1967) claim that the disorder of sound production is central in origin. We endorse the latter theory and believe that the chromosomal defect leads to an as yet unidentifiable morphologic lesion in the CNS manifesting itself in a disorder of cerebral functions and leading to increased neuromuscular excitability of certain centers (Hirschberg, 1974; Schroeder et al. 1967). Indeed, there was no laryngoscopically demonstrable alteration in the patients seen by us. Stroboscopic evaluation is now in course.

KEY POINT

Cri du chat syndrome is a chromosomal abnormality. The outside appearance is characteristic: craniofacial dysmorphism with microcephaly and with somatic and mental retardation. The cry is also very typical, reminding of cat's cry: the phonation is high, shrill, mewlike, and the melody curve is flat. Genetic testing confirms the diagnosis. A satisfactory therapy is not known.

The quality features of the cry are characteristically changed, which may be helpful in establishing the diagnosis by subjective auditive observation and acoustic analysis even prior to chromosome analysis. Genetic testing (FISH) can confirm the diag-

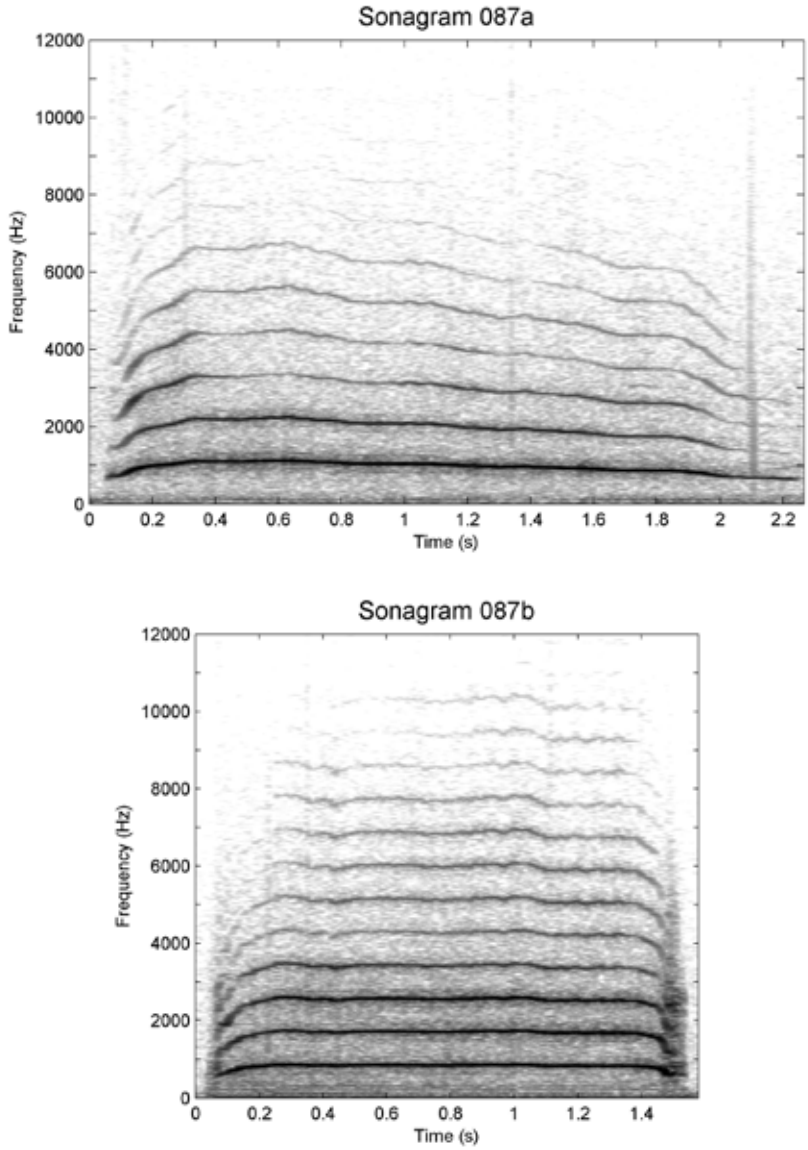


FIGURE 7-74. Cri du chat syndrome. Child aged 2 years. *History:* Thin cry from birth, but no importance has been attached to it. Three months earlier symmetrical paralysis of the upper and lower extremities with paralysis of the diaphragm and cervical muscles, which have improved. Tick encephalitis and poliomyelitis suspected but disproved. *Laryngoscopy:* No pathology. *Acoustic finding:* Cry closely resembling the mewing of a cat pathognomonic for the cri du chat syndrome and not for the intercurrent paralysis. **A**. *Sonagram No. 87a:* Scanty harmonic structure with overtones around 1000, 2000, 3000 to 3300, 4000 to 4500, 5000 to 5600, 6000 to 6500, and so forth. Hz with hundreds of Hz between them. There may be a fundamental of very high frequency, or it may be replaced by an intensive overtone at 700 Hz. Noise components are absent. The above mentioned components are slightly more intensive than the rest. The scanty harmonic structure, in which the overtones show small, irregular undulations and a falling melody in the higher frequency ranges, make the cry shrieking and mewing. The crying phrase may be long (as observed in other parts of the cry), lasting longer than 2 s. The cry is particularly high-pitched due to the intensive component at 1000 Hz. Flat melody. **B**. *Sonagram No 87b:* Mewing of a 1-year-old cat showing striking acoustic similarity to the cry of the cri du chat syndrome infant. Overtones are arranged regularly at grossly 800 Hz intervals above a fundamental of about 800 Hz. First the melody shows a rise of about 100 to 200 Hz, whereas at the end there is a fall of approximately the same value. No noise components are present.

nosis. The dactyloscopic changes, too, may also help in the diagnosis.

No treatment exists for the underlying genetic disorder, so medical care is focused on the symptoms (Chen, 2002).

Myasthenia Gravis

Myasthenia gravis (MG) is an acquired, neuromuscular, autoimmune disease of adults. It is twice as common in women as in men. Its essential feature is a weakness following the use of various muscles, due to a disorder of transmission at the neuromuscular junction. The disorder is characterized by a decrease of the number of acetylcholine receptors in the neuromuscular plates.

MG is uncommon in children but may occur also in neonatal age. For children, an estimate would be 2,050 children with the disease per million (Phillips & Torner, 1996). *It is not necessarily the same condition in children as in adults. One can recognize three separate entities in the childhood period*: (a) transient neonatal myasthenia in an infant of myasthenic mother, (b) congenital or infantile myasthenia in an infant of nonmyasthenic mother, and (c) juvenile myasthenia similar to the adult MG (Dubowitz, 1978; Joshi, Bharucha, Mohire, & Kumta, 1985).

The transient neonatal form develops in children born from mothers with myasthenia. Papazian (1992) reported a 21% incident of transient neonatal myasthenia gravis (TNMG) on infants born to mothers with MG. The condition responds well to treatment and resolves within a few weeks. Complete recovery is expected in less than 2 months in 90% of patients and by 4 months of age in the remaining 10% (Bartoccioni et al., 1986). *In neonates with TMG a faint, weak, inert cry, a forceless phonation may be the first, or, in fact, the only symptom of myasthenia*. Weak cry was reported in 60 to 70% in the neonatal cases (Papazian, 1992). Due to a gradual loss of contractility of the vocal cord muscles, the increasing weakness of the cry is even more noticeable on prolonged crying.

Congenital myasthenia presenting in an infant of nonmyasthenic mother is rare. According to Simpson (1981) in these cases there is no association with thymoma or antibodies, as seen in adult cases. Bundey (1972) suggested an autosomal recessive pattern of inheritance; it is not an autoimmune disease as its adult counterpart. The diagnosis may be difficult and the prognosis is not always good.

Juvenil myasthenia is similar to adult-onset immune related myasthenia gravis except for its age of onset.

Most often, the myasthenic symptom appears in the extraocular muscles: the patient looks permanently sleepy because the eyelids are drooping (ptosis); consequently a double vision may occur. *Involvement of the soft palate, tongue muscles, and esophagus causes disorders of swallowing and speech: rhinophonia aperta and dysarthria* (Hirschberg, 1986; Hirschberg & Gross, 2006). Voice and speech disorders may be one of the leading symptoms in later childhood and in adult age, too (Frint & Nábrády, 1966; Maxwell & Locke, 1969). In the classic case the patient notes progressive swallowing difficulty from the beginning to the end of a meal. The velopalatal weakness leads not only to hypernasality but also to nasal regurgitation as well (Painter, 1983). Muscular fatigue and even breathing difficulty may develop. The symptoms tend to worsen with increased activity and im-

prove with rest. Often, patients feel well in the morning, but as the day goes on, they become weaker.

The first step of diagnosing MG includes a review of history and physical and neurologic examens. If a suspicion of MG exists, *several tests are available to confirm diagnosis*. They include: antibody blood test, nerve conduction test/repetitive stimulation, single fiber electromyography, CT or MRI for identifying an abnormal thymus, and most commonly the Tensilon (edrophonium chloride) test.

KEY POINT

Myasthenia gravis is an acquired, neuromuscular, autoimmune disease: it is a disorder of the transmission at the neuromuscular junction. In infants it is not necessarily the same condition as in adults (not an autoimmune disease) and may have three different types. Faint, weak, inert cry is characteristic which clears up and becomes powerful following i.v. Tensilon. If the soft palate is also involved, hypernasality of the voice may occur. Several tests are available to confirm the diagnosis, among others the stapedius reflex test. The therapy is individually determined with respect to the age of the patient.

Using the latter, when this drug is injected, the weak eye muscles of the patient briefly get stronger, and in infants the weak cry clears up, and becomes forceful and regular (Figures 7-75A and B). The improvement of the speech of older children with MG may be sensational in the course of intravenous administration of Tensilon while counting from 1 to 40: the hypernasality decreases gradually and at the end of counting the speech will become totally clear and forceful (see attached DVD). As the MG also affects the smallest muscle of the human body, the stapedius, the stapedius (acoustic) reflex test also may be applied, using the Tensilon probe. After the injection the change of the curve will be evident (Figures 7-76A and B). Electromyography also may be used for to differentiate between myogenic and neurogenic processes (Figure 7-77).

For differential diagnosis of the neonatal and juvenile forms of MG one can find an outstanding guide in the article of Brueton et al. (2000).

The therapy should be individual, and planned with respect to age. The most applied methods are the following: pyridostigmine (Mestinon) is the usual first-line treatment for MG. Infants with severe weakness from transient neonatal myasthenia may be treated with oral pyridostigmine, but mechanical respiratory ventilation also may be necessary. Prednisone is used for long-term immunosuppression; it is the most effective treatment for ocular MG. Plasma exchange and human immune globulin are used when MG patients have life-threatening signs such as respiratory insufficiency or dysphagia. Thymectomy is performed for long-term benefit in patients aged 8 to 55 years with generalized MG. The prognosis depends on the type of the disease. In general, the long-term prognosis for the patient with congenital myasthenia is changing: most patients remain fairly stable throughout their lifetime and tend not to have wide fluctuations of symptoms or function nor myasthenic crisis. The future of MG lies in the elucidation of the molecular immunology of the antiacetylcholine receptor response with the goal of developing a rational treatment for the illness that will

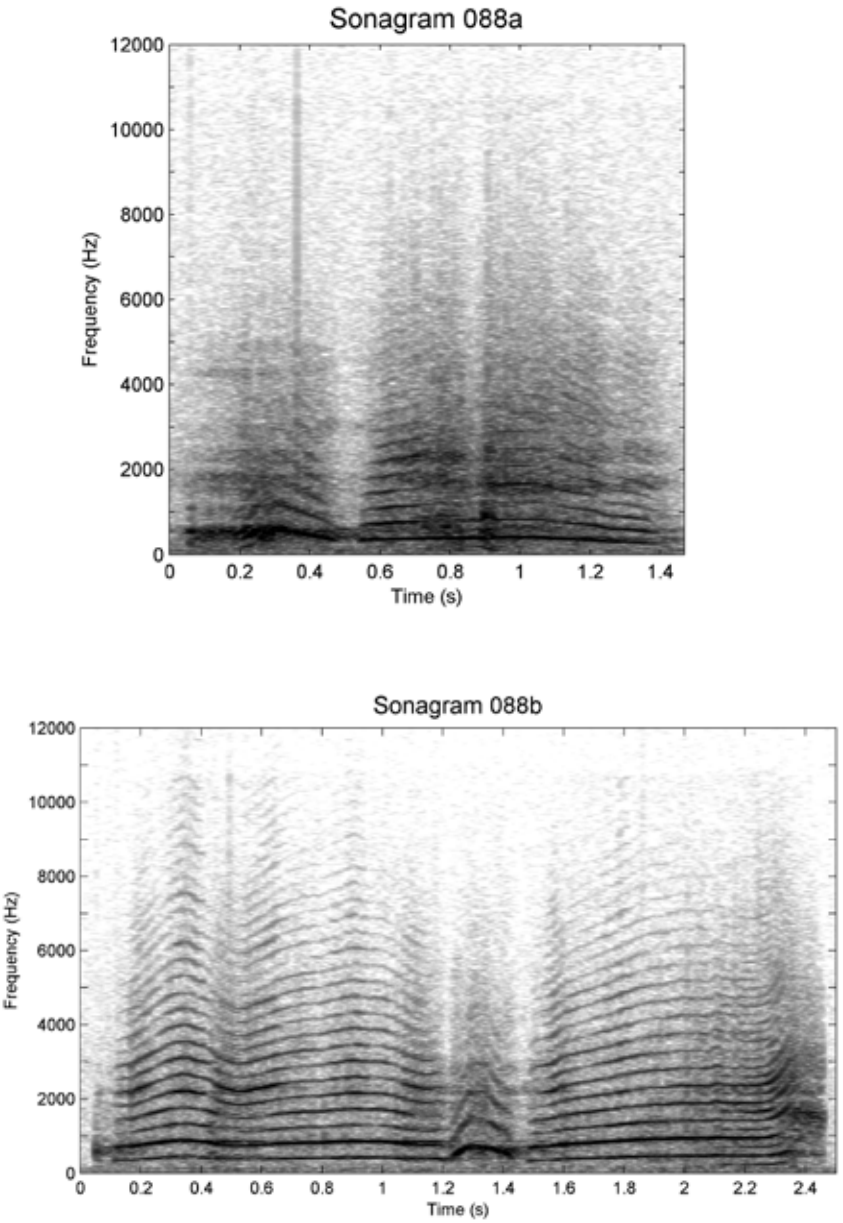


FIGURE 7-75. Myasthenia gravis. Child aged 23 months. *History:* Slightly "hoarse" cry from birth. Treated in hospital for bronchitis at 4 months of age. Unable to open his eyes properly since. Difficulty of swallowing. *EEG:* No convulsive potentials. *Radiography of skull:* Normal. *Laryngoscopy:* No pathology. *Acoustic finding:* Faint, inert, and veiled cry (**A**), which clears up and becomes powerful following i.v. Tensilon (**B**). **A**. *Sonagram No. 88a:* Regular harmonic structure, but the overtones around 750, 1500, 2100, and 2800 Hz are much more intensive than those below 1500 Hz, presumably because the former are strongly masked by noise. Massive masking by noise from 1500 Hz up to the top of the spectrum, some parts showing an overtonelike arrangement. **B**. *Sonagram No. 88b:* Regular harmonic structure with the overtones about 400 Hz apart. Some overtones, especially those between 2000 and 3000 Hz, are thickened. The cry is intensified and becomes normal and slightly presssed (from 2.25 to 2.45 s slightly hollow) in response to Tensilon administration.

A B

FIGURE 7-76. Myasthenia gravis. Child aged 2 years. Stapedius reflex test (**A**) before and (**B**) after i.v. Tensilon injection.

cure the abnormality in the immune system (Howard, 1982, 1990, 1997; Sanders & Howard, 1995).

Amyotonia Congenita (congenital muscular atony, Oppenheim's disease)

The disease is a rare congenital, noninherited but sometimes familial disorder and is characterized by hyperflexibility of the joints, and atony and hypokinesis of the muscles (triad of Oppenheim). The disease was first described by Oppenheim (1900).

KEY POINT

Congenital amyotony is a noninherited but sometimes familial anomaly. It is characterized by hypotonia of the muscles, including vocal cords, with the consequence of a wan cry. The coughing sound is also inert and weak. The diagnosis may be confirmed by electromyography.

The symptoms which can be observed soon after birth, include a conspicuous flaccidity and weakness of the muscles and an unusually great passive mobility of the extremities. Owing to the excessive decrease of muscle power, the extremities appear to be totally paralyzed. The etiology is unknown. It has been suggested that changes of the motor endplates lead to the clinical symptoms (Horányi, 1962).

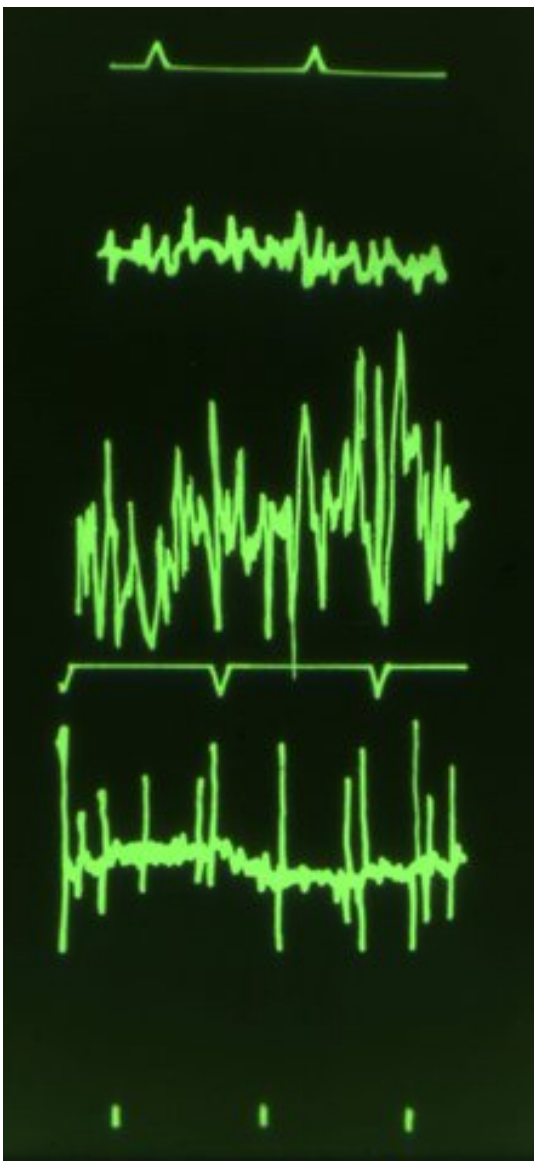


FIGURE 7-77. Electromyograms performed on the levator veli palatine muscle: (**A**) myogen disorder, (**B**) normal electromyogram, (**C**) neurogen process.

The concomitant changes in phonation are also characterized by hypotonia: the cry is wan (Figure 7-78), and the cough is inert and weak.

The diagnosis, postulated on grounds of the above symptoms, can be confirmed by EMG and muscle biopsy.

Polyradiculitis (Guillain-Barré syndrome)

Acute polyradiculitis is an acute inflammatory disease of the peripheral nervous system (Grisold, Drlicek, & Liszka, 1991). *The pathologic changes develop mainly in the spinal nerve roots*; many spinal nerve roots may be inflamed. This condition may occur in several diseases, the Guillain-Barré syndrome (named also radiculoneuritis or neuronitis) is a special form of this disease. According to Horányi (1962) this condition is, in fact, histologically not inflammatory but a degenerative process, involving the myelin sheaths and axons. It mostly occurs in conjunction with infectious diseases (diphtheria, influenza) and is due to the effect on peripheral nerves of the endotoxins or exotoxins of the pathogenic agents. A general muscular weakness develops with paresthesia and pain, the patient is unable to walk, then symmetric paralysis develops in other muscles.

In the acute phase, respiratory insufficiency and autonomous dysfunction may occur. When the muscles of the trunk and the respiratory muscles are also involved, characteristic changes can be noted in sound production, resulting in the weakness of the cry and cough (Figures 7-79A and B).

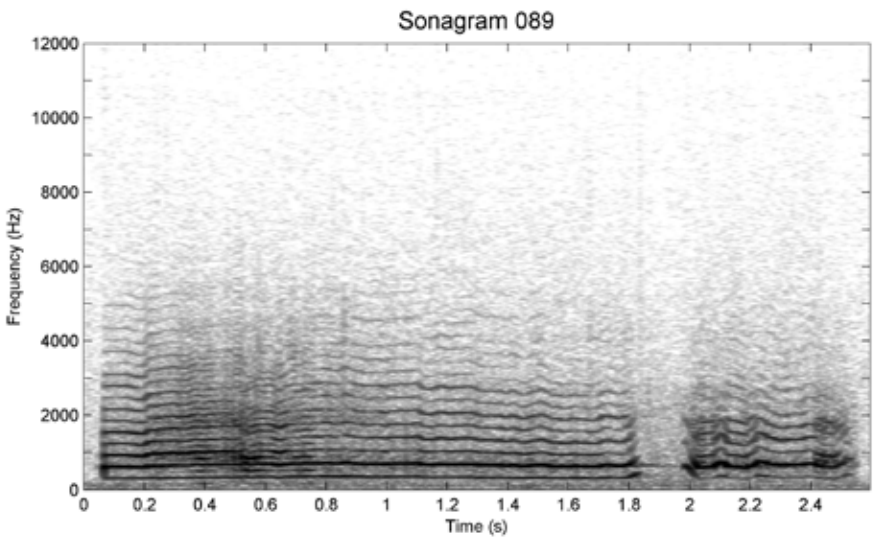


FIGURE 7-78. Congenital amyotony (Oppenheim's disease). Infant aged 2 months. *History:* Poor mobility and inert, hoarse cry from birth. Sleeps a lot, gets tired on feeding. Holds the right arm abnormally. Sibling died of "muscular weakness." *Neurologic examination:* All four extremities are hypotonic; trunk muscles and hands in forced position; thighs rotated outward. Blue sclera. *Electromyography:* Unreliable, no conclusion can be drawn as regards the central or peripheral origin of the disease. *Laryngoscopy:* Vocal cords mobile, no paresis, diameter of laryngeal lumen normal. *Acoustic finding:* Low, faint, and wan cry and a very weak cough.
Sonagram No. 89: Up to 2800 Hz apparently normal harmonic structure; scanty information above this frequency. Occasionally the cry is interrupted and becomes undulating presumably because of imperfect innervation. In average free of noise. The cry has a regular ring, but it is a low-energy sound.

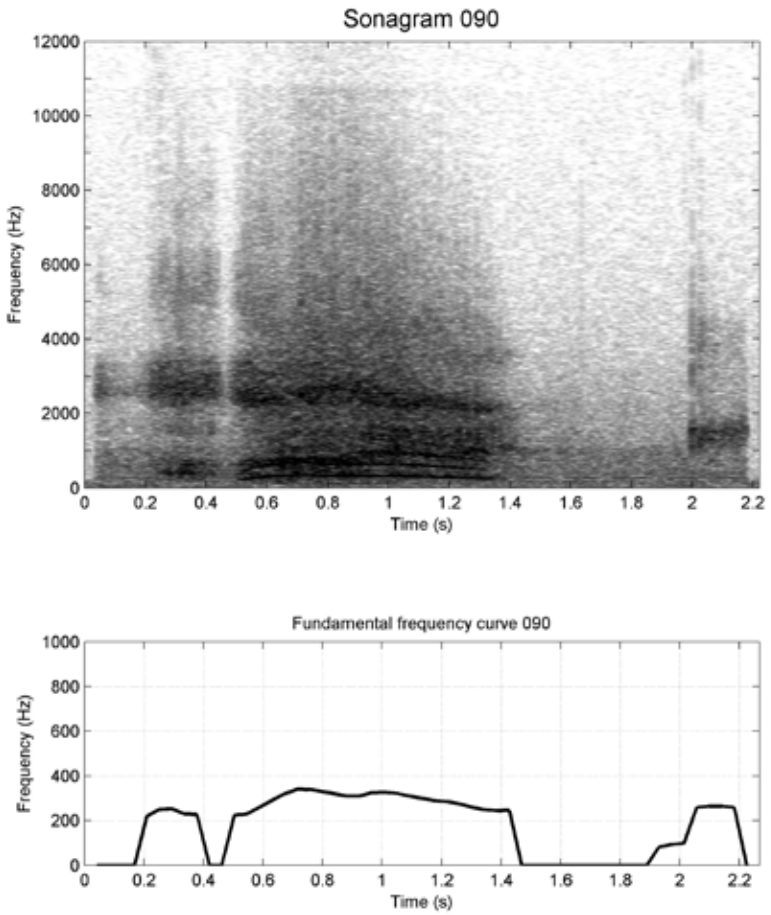


FIGURE 7-79. Polyradiculitis. Child aged 30 months. *History:* Common cold for 2 weeks; high fever, discharge from the ears for 4 days; crying constantly, hoarseness and wailing for 3 days, unsteady gait, restricted mobility of the left hand for 2 days. *On admission:* Reduced muscle tone, unable to lift its legs and arms, grasping power reduced, tendon jerks cannot be elicited. *Laryngoscopy:* The vocal cords are intact; they seem to close during phonation and show excursions during inspiration. Hypofunctional dysphonia of central origin. *Acoustic finding:* Hypotonic, faint, inert phonation and wailing. **A**. *Sonagram No. 90:* Low-intensity fundamental frequency without a clear harmonic structure above. Instead, occasional broad, blurred, high-intensity bands appear between 2500 and 5500 Hz. Strong masking by noise, which might be explained by conscious complementary noise generation compensating for the weak phonation. The preponderance of the components above 2500 Hz frequency makes the voice faint. **B**. In the *fundamental frequency curve*, which integrates the acoustic components all over the spectrum. The fundamental is in the high-frequency band, as the relative intensity of the lower frequency components is low and is

Children who survive more then 18 months after the onset of the symptoms may show slow improvement. According to Steiner and Abramsky (1985), it is possible that immunologic, and immunogenetic, as well as infective, environmental, and other factors, must be present to facilitate the development of peripheral nerve damage, but the possible immune pathogenesis has not yet been proven. Recently Gazzola et al. (2001) reported that possibly hepatitis C virus is involved in acute meningoradiculitis/polyradiculitis of HIV1 coinfected patients.

For diagnosis, predominantly clinical criteria are used according to the criteria summarized by Grisold, Drlicek, and Liszka, (1991).

Even though there are several procedures of treatment that generally reduce the duration of the disorders, a standard and reliable cure is not available. Recovery is spontaneous and therefore impossible to predict. Relapses are rare, but may occur many years later.

In polyradiculitis type Guillain-Barré early cortisone therapy is recommended, administration of immunoglobulin also may be helpful. Therapeutic local anesthesia (continuous nerve blockades) influences the pain advantageously. Despite extremely severe courses and complications the prognosis is favorable for the majority of patients.

KEY POINT

Polyradiculitis is an inflammatory (or degenerative?) disease of the peripheral nervous system, in particular, of the spinal nerve roots. A general muscular weakness develops eventually resulting in the weakness of the cry and cough. In the acute phase cortisone and immune therapy may be helpful.

Pseudocroup (subglottic laryngitis)

Pseudocroup is a disease occurring in countries with a moderate climate, mainly in the fall and early winter months. Older infants and younger children are affected. The onset is almost invariably marked by a "grippe" (virus) infection. It is caused more frequently by parainfluenza virus, rarely by adenovirus, ECHO virus, or influenza virus. Usually at night, the child, who has had a common cold, catarrh, and low fever for a few days, is wakened by a bout of cough, usually associated with a "drawing" inspiration. Episodes of dyspnea last several hours and resolve spontaneously. Dyspnea may recur in the same night or thorough several subsequent nights. It is impossible to forsee the degree of the episodes and their frequency (Chmielik & Kaczmarczyk, 2006).

The symptoms are due to a bilateral, symmetric, cushionlike, pale pink swelling of the mucosa in the subglottic area, the conus elasticus: Figure 7-80. The vocal cords are not affected or merely slightly inflamed, and the changes in the lower airways (trachea, bronchi) are those of a slight catarrh, at most. Thus, essentially the process is limited to the subglottic area. This is what differentiates it from the much more serious, stenosing, malignant laryngotracheitis, in which marked changes occur in the larynx and trachea, in addition to those in the subglottic area. The two conditions are discussed as a single entity in several textbooks (Biesalski & Collo, 1991; Ferguson & Kendig, 1972). We think, however, that a differentiation is justified pathogenetically, and morphologically as well as for reasons of education.

The sound phenomena produced in pseudocroup are typical and are obviously results of the changes demonstrable by laryngoscopy. The vocal cords are intact; therefore, the *cry is clear, or only slightly veiled*, but never hoarse; sometimes it may be hollow or dull but is always free of noise. *The airway obstruction caused by the subglottic swelling produces an inspiratory stridor which is subglottic in character, hollow, or deephollow. The inspiratory phase is usually protracted, and the respiratory rate high. The cough caused by the catarrh*

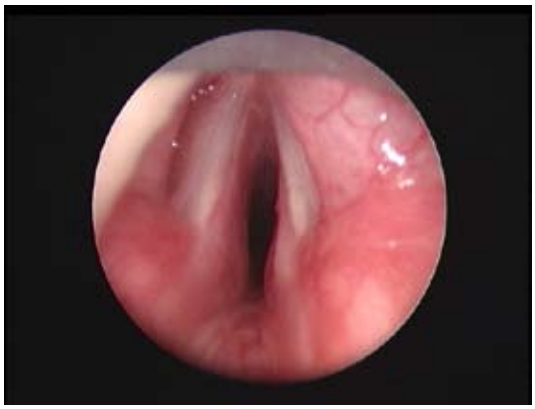


FIGURE 7-80. Laryngitis subglottica (pseudocroup).

is hollow or barking, due to the subglottic change. The typical sound triad, clear cry, inspiratory stridor, and barking cough, are shown in Figures 7-81A and B. In severe cases with excessive obstruction, there may be jugular or epigastric retraction and also cyanosis. In such cases the infant is restless and frightened.

KEY POINT

Pseudocroup is the consequence of a viral infection in older infants and younger children. The beginning of the disease is typical: the fewerish, catarrhal child awakes in the night by a bout of cough and a "drawing" inspiration. The presence of the characteristic sound triad (clear cry, hollow stridor, barking cough) may help in orientation. Calming and cold humidification are the first steps to be taken. Severe cases needs hospitalization where the diagnosis can be unambiguously established by laryngoscopy. The laryngoscopic picture is typical: bilateral, symmetric, cushionlike pale pink swelling may be seen in the subglottis. Corticosteroids, racemic epinephrine, short intubation may be necessary in the management.

In general, children presenting with symptoms of pseudocroup have a milder degree of airway obstruction and in these cases the patient can be treated as an outpatient with supportive measures such as cold humidification. One has to stress, however, that *a certain and final diagnosis can be made only by laryngoscopy.* Thus, in uncertain cases, especially, small children should be hospitalized. On occasion, these patients may have more significant airway involvement, necessitating the use of corticosteroids, racemic epinephrine treatments, and careful observation by experienced personal (Sie, 2000).

Systematic antiinflamatory drugs, and humidification in the room is recommended. In severe cases short-time intubation also may be performed which protects the child from asphyxiation. If the infant is younger than 6 months, one must always think of the possibility of a congenital subglottic stenosis or subglottic hemangioma.

Subglottic Stenosis

Subglottic stenosis (SGS) may be congenital or acquired. The congenital subglottic stenosis is the third most common laryngeal anomaly (Myer & Cotton, 1989), in our patient base the second one (Hirschberg, 1989). The SGS is more often an acquired alteration due to intubation; see previous section (Linna et al., 2004). The congenital laryngeal stenosis is mostly limited to the subglottic area (Hirschberg, 1989). It may produce severe neonatal asphyxia, yet the clinical symptoms are usually not so dramatic as they are in the case of laryngeal atresia (see previous discussion).

As for the severity of the obstruction (subglottic and laryngotracheal steno-

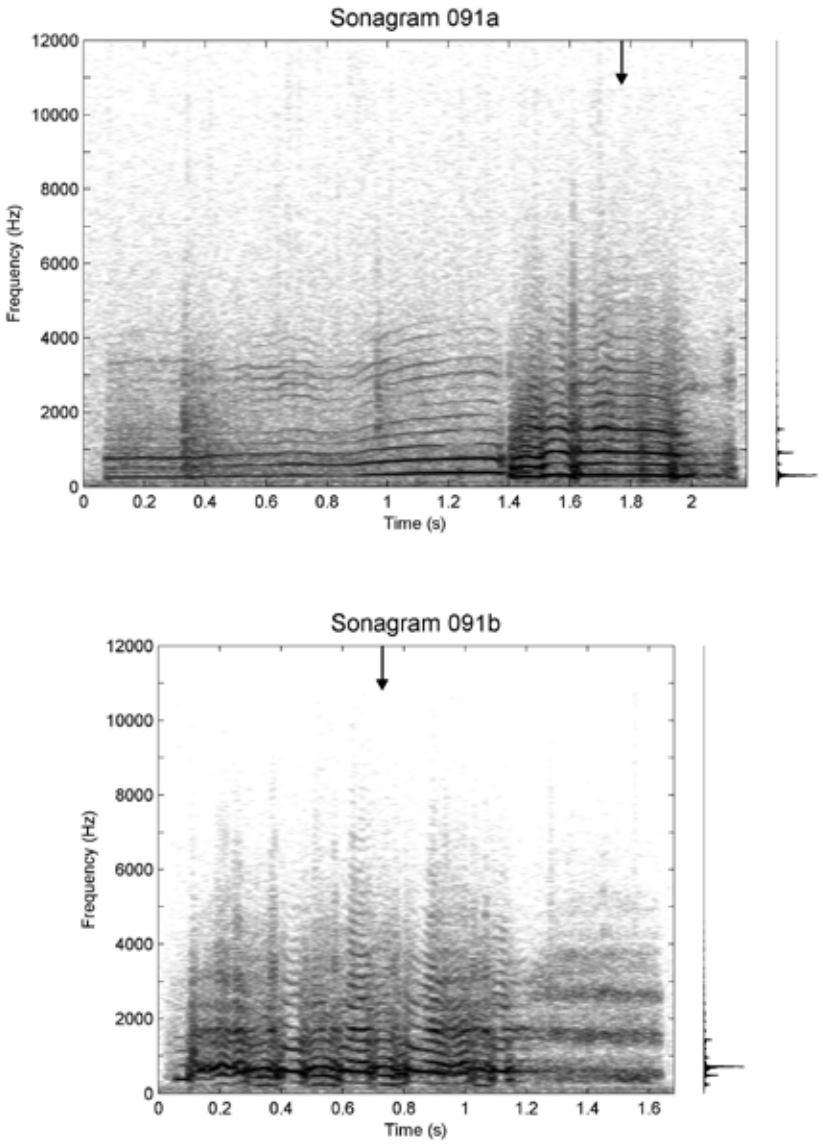


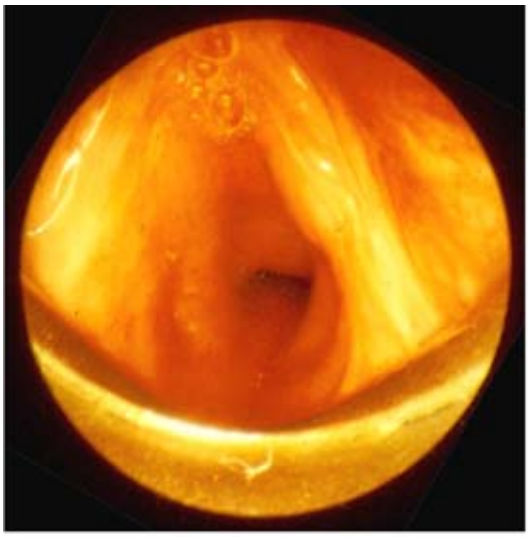
FIGURE 7-81. Pseudocroup (subglottic laryngitis). Child aged 2 years. *History:* Common cold with catarrh for a few days. "Drawing" cough, "hoarseness," and restlessness at night. *Laryngoscopy:* Typical cushionlike bulges on both sides in the subglottis, which is free of inflammation. *Acoustic finding:* Dull, but clear cry; deep subglottic inspiratory stridor (A); hollow, barking cough (B). **A**. *Sonagram No. 91a:* 0.05 to 1.15 s: Regular, intensive fundamental frequency and a strongly defective harmonic structure with only a few components over 1200 Hz, which appear intermittently for a short time. No noise. The absence of harmonic structure above 1200 Hz makes the otherwise clear cry typically dull; 1.2 to 1.65 s: Regular harmonic structure up to 5500 Hz; the overtones show characteristic undulations at about 0.05 s intervals. The single overtones are frayed and thickened, but less so at higher frequencies. Due to the predominance of the fundamental and of low-frequency overtones, the stridor appears to be low pitched. **B**. *Sonagram No. 91b:* During the entire length of the sound phenomenon a densely ribbed harmonic structure is visible. This falls into an explosive phase with a maximum frequency of 5000 Hz and a short phase, where the harmonic structure is complete. Intensive noise bands especially between 500 and 700, 1600 and 1900, and 3300 and 3600 Hz; they become weak or disappear during the second phase of the cough. The pulsating character of the cough is due to the alternation of noisy and clear phases. As a result, the cough is both hollow (densely ribbed) and barking.

sis), Cotton (1991) and Myer, O'Connor, and Cotton (1994) proposed a four-grade classification: (I) means 0 to 50% stenosis, (II) 51 to 70% stenosis, (III) 71 to 99% stenosis of the lumen, and (IV) total obstruction. According to another classification (Lee, 2006), the size of the actual tube that fits the airway is compared with the expected tube size, and the percent obstruction is assigned a grade level.

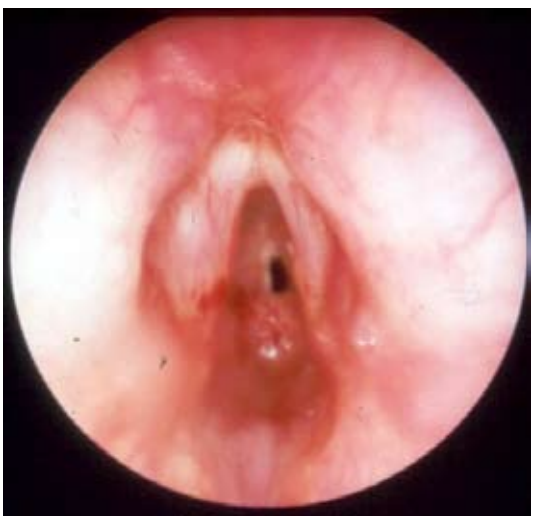
KEY POINT

The typical sound triad commonly characteristic of any kind of subglottic stenosis consists of: (1) clear cry, (2) hollow inspiratory stridor, and (3) barking cough. Causing factor and etiology can be established only with direct laryngoscopy.

The cause of *congenital subglottic stenosis* is in utero malformation of the cricoid cartilage (McClay, 2006). In the apprehension of Ferguson (1972a) it is a defective development of the cricoid cartilage or the conus elasticus. As is well-known, the subglottic airway is the narrowest area of the airway: in normal neonates the lumen of the cricoid cartilage is 5 to 6 mm in diameter, in premature infants 4 mm. If it is narrower (3.5 mm or less), or the cricoid cartilage is excentrically shaped or positioned, we speak of subglottic stenosis. In other words: one may speak about a SGS when a scope which corresponds to the age of the infant, cannot be passed through the stenotic area. Congenital stenosis is never membranelike, it is mostly diffuse (Figure 7-82A), rigid and free of edema or inflammation. Incidentally, trisomy 21 is associated with a higher incidence of this anomaly.



A



B

FIGURE 7-82. Endophoto about a congenital (A) and an acquired (B) subglottic stenosis. Child aged 5, and 11 months, respectively.

Though the respiratory disorder is mostly congenital, the symptoms present themselves in the first few months of life rather than at birth and they may remain latent until a respiratory infection (recurrent pseudocroup) or intubation cause swelling of the mucosa and severe obstruction. *A typical inspiratory or biphasic stridor* demon-

strated in 40% of the 84 cases of Fearon and Ellis (1971), *which may also be hollow in character, is the most important acoustic feature of this stenosis.* In the presence of catarrhal or inflammatory components the stridor may be accompanied by a hollow cough (a permanent cough occurred in 20% of the 84 cases of Fearon & Ellis). *The cry is clear, strong, and occasionally hollow* (Figure 7-83).

Acquired subglottic stenosis is the most common acquired anomaly of the larynx in children and the most common abnormality requiring tracheotomy in children younger than 1 year (McClay, 2006). Early in the 20th century, it was usually related to trauma or infection (syphilis, tuberculosis, diphteria). In the late 1960s it occurred in increasing number due to long-term intubation, introduced by McDonald and Stocks (1965). Mechanical trauma from an endotracheal tube can lead to mucosal edema and hyperemia. These conditions then can progress to pressure necrosis of the mucosa. Later, infection of the perichondrium may result in a subglottic scar causing stenosis (Figure 7-82B). Congenital anomalies also

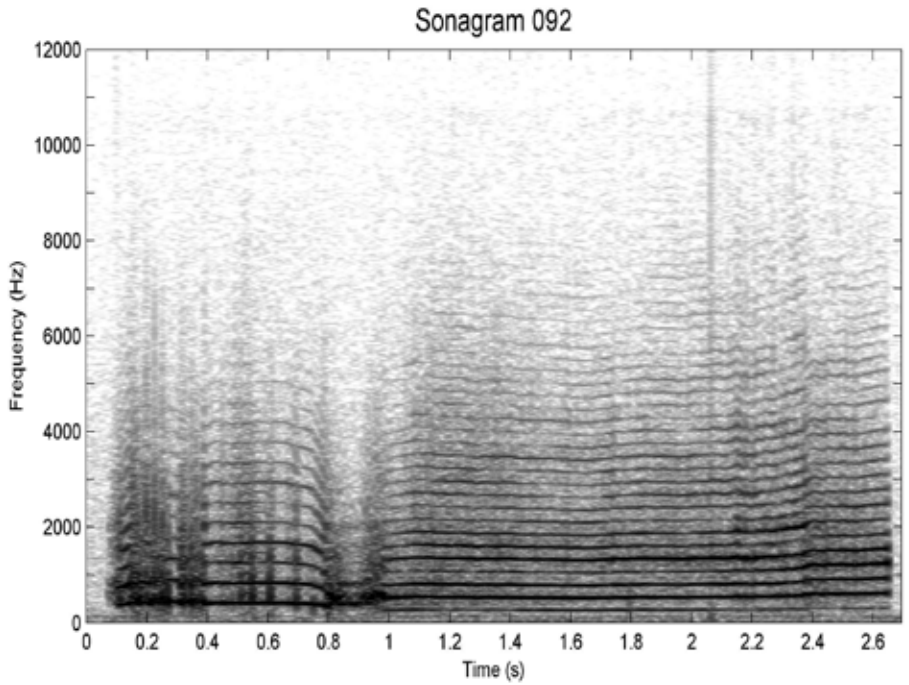


FIGURE 7-83. Congenital subglottic stenosis with secondary chronic subglottic laryngitis. Infant aged 14 months. *History:* Repeatedly treated in hospital for subglottic laryngitis. Has been going to the crèche for half a year; falls ill within 1 or 2 days when among children. Fever for 4 days with suffocating, spastic paroxysms of cough at night. *Laryngoscopy:* Subglottic swelling, stenosis (mainly on the sides and in the back) reducing the lumen to half of its normal size. Tracheotomy and laryngofission with excision of the thickened mucosa below the vocal cords had to be performed (laryngotracheal reconstruction). *Acoustic finding:* Hollow stridor of subglottic character; hollow cry; occasional barking cough. *Sonagram No. 92:* 0.15 to 0.85 s: Scanty harmonic structure up to 5500 Hz, dense ribbing at onset. Fraying of some overtones. Typical subglottic stridor which is hollow at first; 0.9 to 2.4 s: Regular harmonic structure, then, without interruption, two marked phases of dense ribbing (1.25 and 1.65 s). Masking by noise throughout. Hollow cry.

may have a role in the development of acquired subglottic stenosis; it may arise or may be manifested as a result of mechanical trauma, mostly caused by prolonged ventilation (see previous discussion). The number of occurrences of acquired subglottic stenosis has greatly decreased over the past 40 years from 24% to 12% after introducing very small endotracheal tubes with a 2.5-mm internal diameter (Wallner, Ouanounou, Donelly, & Cotton, 2000).

The diagnosis can be made by endoscopy. According to Baker, Kelchner, Weinrich, Lee, Willging, Cotton, et al. (2006) *definitive diagnosis of SGS is made via direct laryngoscopy and bronchoscopy under anesthesia.* During flexible fiberoptic laryngoscopy, topical anaesthesia and decongestion can be accomplished in older infants and children with topical Afrin and lidocaine. If the child is older, videostroboscopy can be performed to assess the vocal function. We agree with McClay (2006) in that, occasionally, the subglottis can be visualized with flexible endoscopy. However, rigid laryngoscopy and bronchoscopy are the safest procedures and offer the best visualization for the subglottis and tracheobronchial fee. In the practice of the Budapest team (Lellei, 2004) various methods are applied; see Chapter 5. Nowadays mostly two methods are used by us on the basis of experiments with 314 direct laryngoscopic investigation in the last 10 years (1995–2004): (1) Application of short Hopkins optic after revealing the larynx with a McIntosh instrument, or (2) Use of a small fiberoscope which may be introduced through a laryngeal masque. With these methods the continuous oxygen apply and the respiratory support are guaranted. According to Yavascaoglu, Tokat, Mogol Basagan, Kaya, Erisen, and Kutlay (2001) the laryngeal mask airway may be superior to tracheal intubation or the use of a face mask during anaesthesia management in severe subglottic stenosis. Laryngoscoppy and bronchoscopy is suitably supplemented with various light sources, video documentation equipment, and telescopes. Certain radiographic examination can help in obtaining the diagnosis, such as anteroposterior and lateral plain neck radiography or fluoroscopy. Laryngotracheography may outline the length of the stenosis (see Figure 7-60). CT scans and MRIs are not often used in the primary evaluation of subglottic stenosis.

The therapy is determined by the measure of the obstruction. *The four forms of treatment* are the following:

1. careful observation and waiting for spontaneous improvement of the respiratory symptoms,
2. medical therapy,
3. endoscopic procedures,
4. surgical methods.

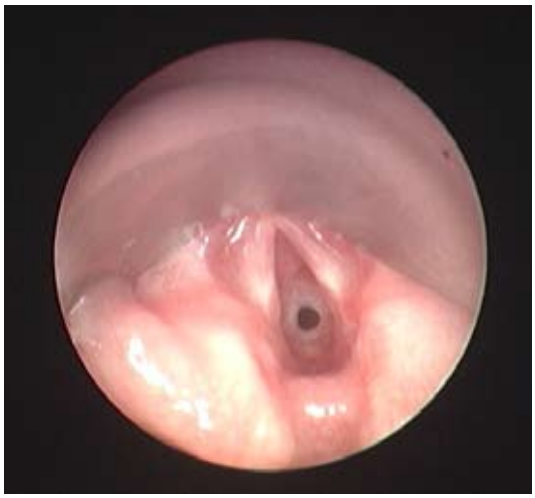
In mild cases observation is necessary; with advancement of the age the symptoms generally subside with relative enlargement of the airway lumen and may eventually cease entirely without intervention. No known medical therapy for mature subglottic stenosis exists (McClay, 2006). Treatment of the inflammatory process with oral or inhaled steroids can sometimes decrease the severity of the disease. For mild or granular stenosis, investigators have reported success with serial endoscopic dilatation with or with-

out steroid injection. Carbon dioxid laser was also an option for soft circumferential stenosis, but normally dilatation and the use of a laser may cause scars and recurrence of the stenosis. In addition, topical application of mitomycin C (MMC) has been recommended to inhibit scar formation: Simpson and James (2006) suggest that MMC is an effective adjuvant in the treatment of laryngotracheal stenosis. A control study (Hueman & Simpson, 2005) has shown, however, that there is no benefit of its use in mature stenosis. Eliasar, Gross, Maly, and Sichel (2004) also stress on the basis of an animal study that mitomycin does not prevent laryngotracheal repeat stenosis after endoscopic dilatation surgery. In severe cases tracheostomy is unavoidable and a decannulation in these patients is possible only with a larynx widening operation. Besides, the mechanical endoscopic dilatation electrocauterisation, cryosurgery, and several stents had been used until the 1970s; restenosis was, however, a frequent consequence, in about 50% of the cases.

Since then *two main types of external surgical solutions have been applied for children*. One is the *incision technique with laryngotracheal reconstruction* and postoperative stenting (Cotton, 1978, 1991, 1995, 2000; Cotton & Evans, 1981; Cotton & Myer, 1998; Cotton, Myer 3rd, O'Connor, & Smith, 1995; Cotton & Seid, 1980; Cotton & O'Connor, 1995; Evans & Todd, 1974; Fearon & Cotton, 1972; Fearon, Crysdale, & Bird, 1972; Seid, Pransky, & Kearns, 1991; Zalzal & Cotton, 1986). These various procedures may be divided into three groups. The first is anterior laryngofissure, with or without cartilage grafts. The second is anterior laryngofissure and posterior division of the cricoid. Four quadrant division of the cricoid may also be performed. Evans and Todd's (1974) initial procedure divided the upper trachea and cricoid in a castellated incision ("stepped incision") to allow expansion. Later Cotton (1978, 1991) described a technique of anterior division along with insertion of a cartilage graft without a stent. According to Koltai, Ellis, Chan, and Calabro (2006) the dimensions of cartilage grafts are important in successful laryngotracheal reconstructions. In their 54 cases rib cartilage was used in 51 patients and thyroid cartilage in 3 patients. Forty- eight (89%) of them were successfully decannulated. The use of 2-mm increments for the posterior graft suggests a set of molds that are 2, 4, and 6 mm wide and 22 mm long. Using 2 x 2-mm increments for the anterior grafts indicates that 36 mold sizes will be sufficient for 90% of predicted cases. Younis, Lazar, and Bustillo (2004) conclude that laryngotracheal reconstruction with a costal cartilage rib graft should be considered the procedure of choice for the management of subglottic stenosis. In the Heim Pál and Budai Children's Hospital (Budapest) we have applied the modified Réthi–Bánfai (Réthi 1956, 1959) method: decannulation was possible in all 42 children (Lellei, 2004; Lellei & Hirschberg, 1983, 1988).

Figures 7-84A through E show the surgical procedure in a 1-year-old baby with subglottic stenosis (Grade 3). In this case an anterior rib cartilage graft was applied by us, seen also on the drawing.

The other surgical technique is the *cricotracheal resection* (CTR): excision of the stenotic area and an end-to-end anasto-



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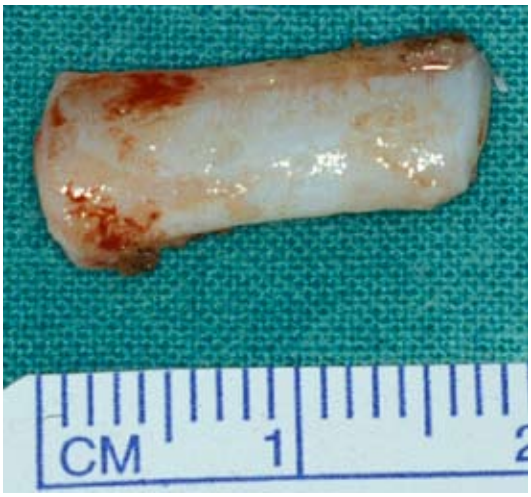

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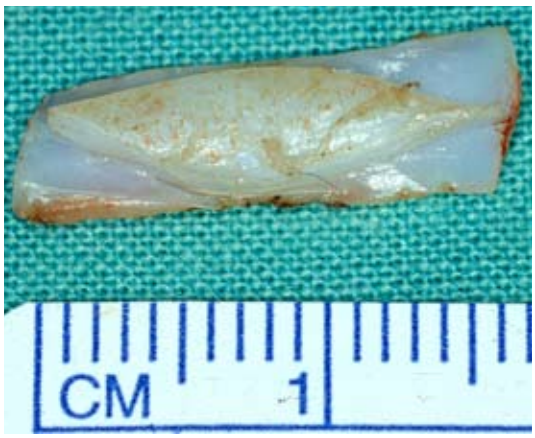

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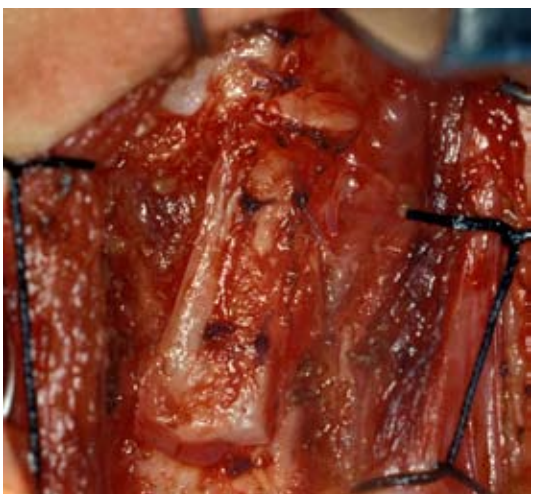

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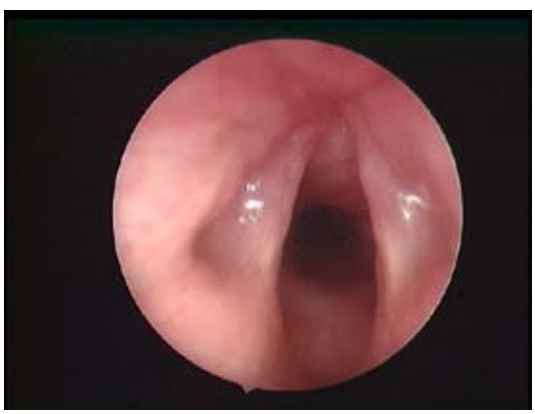
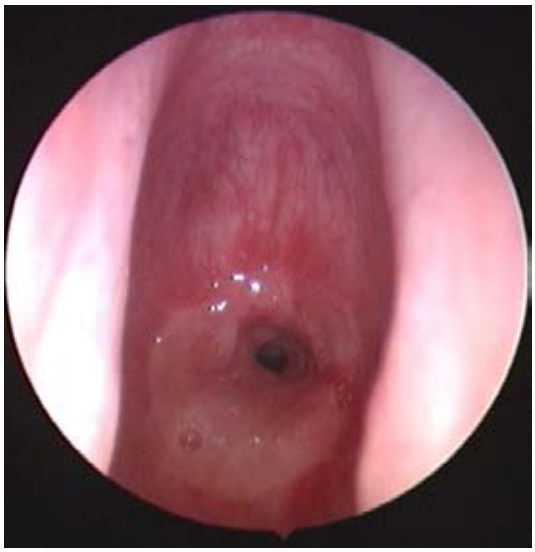


E

FIGURE 7-84. A. Subglottic stenosis, grade 3. Infant aged 1 year. **B**. Rib cartilage; **C**. subglottic stenosis carved anterior cartilage graft; **D**. graft in place; **E**. 3 years postop.



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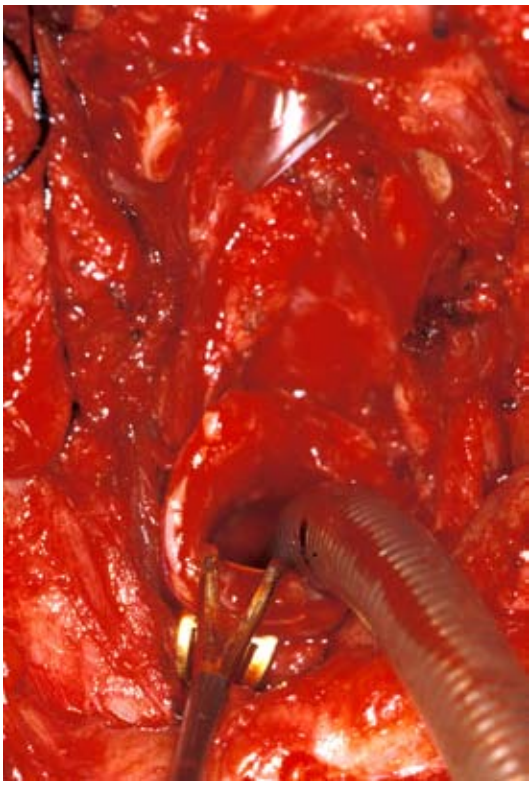

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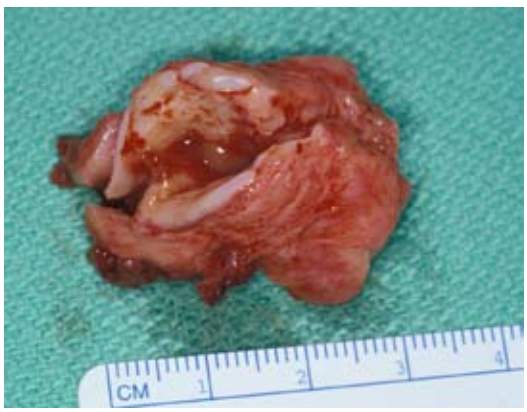

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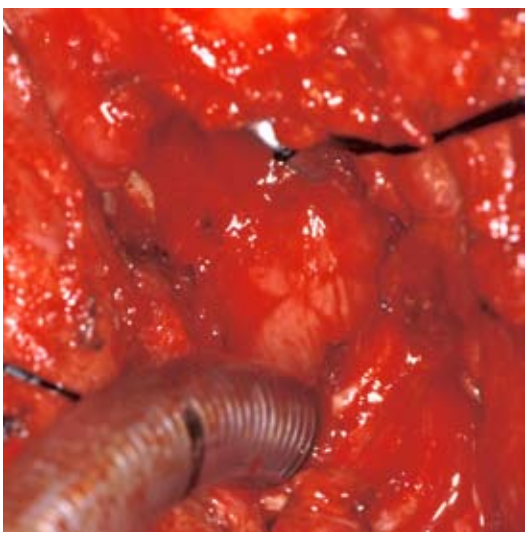

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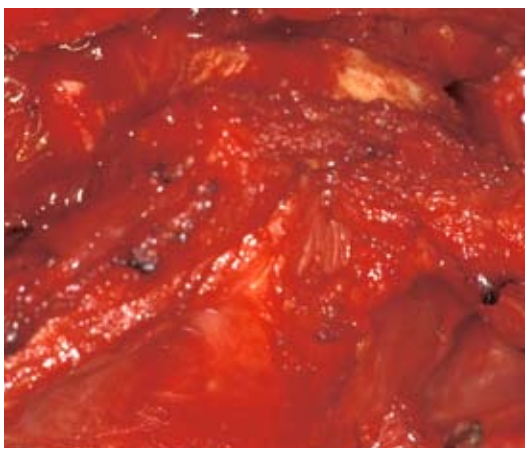

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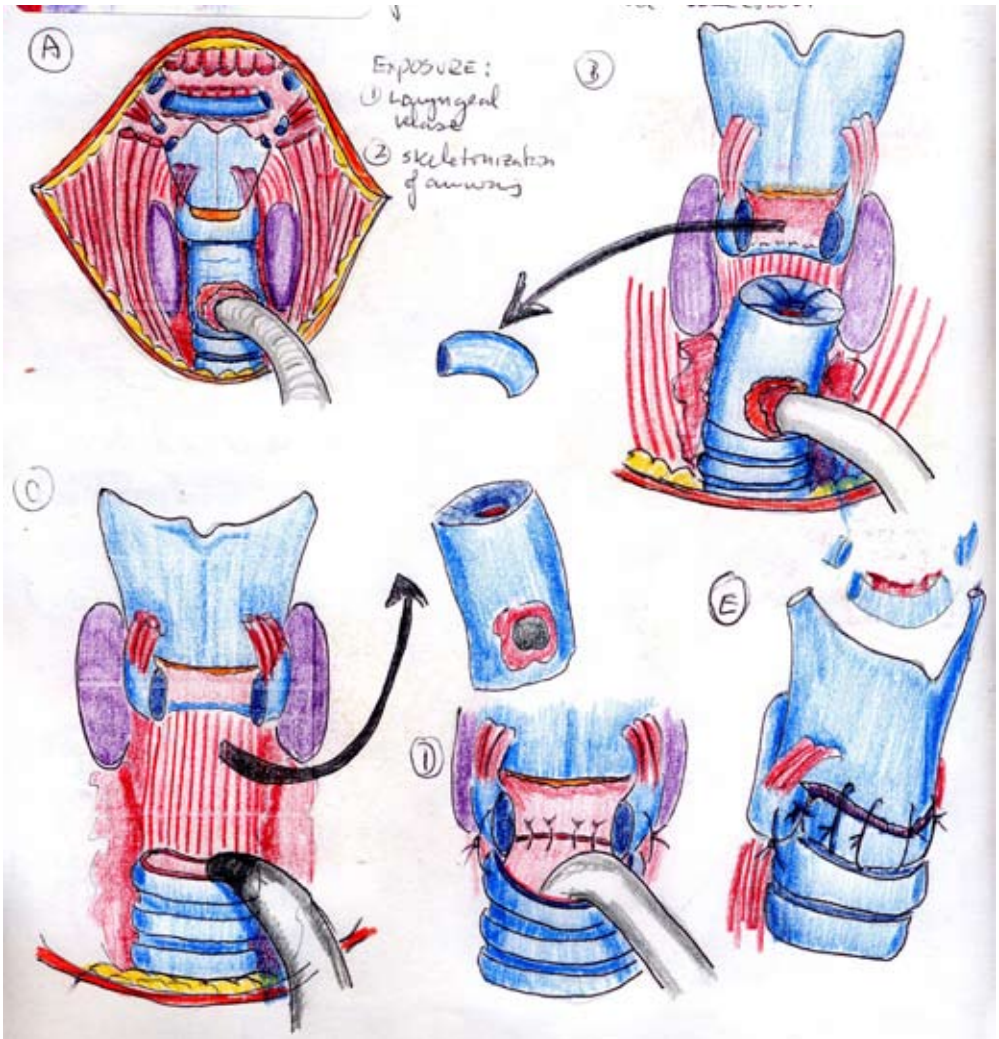

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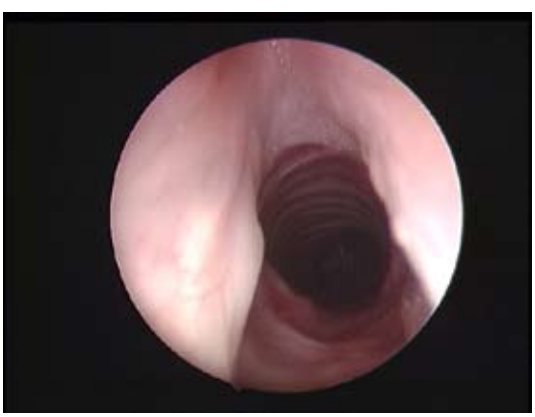


G

FIGURE 7-85. **A**. Circumferential subglottic stenosis; **B**. cricotracheal resection. Anterior cricoid and 4 tracheal rings resected. **C**. Specimens; **D**. posterior anastomosis; **E**. anterior anastomosis; **F**. 4 months postcricotracheal resection. **G**. Steps of surgery: drawings (Koltai).

mosis (Bailey, Hoeve, & Monnier, 2003; Hartley & Cotton, 2000; Walner, Stern, and Cotton, 1999). In Hungary Czigner (2004) managed combined subglottic and tracheal stenosis by a one-staged operation: circumferential resection of the subglottis and trachea with primary end-to-end anastomosis in 23 patients: 21 among them are now permanently extubated. Cricotracheal resection provides a surgical means to treat high subglottic stenosis that closely approximates the true vocal cords—according to Boseley and Hartnick (2006).

Figures 7-85A through G show the surgery of a circumferential severe subglottic stenosis solved by cricotracheal resection: anterior cricoid and 4 tracheal rings were resected.

The basic question asked by Hartley and Cotton (2000): is that laryngotracheal reconstruction or cricotracheal resection? In general, we can say with Lee (2006) that the goal of any laryngotracheal surgical reconstruction is decannulation and re-establishment of the airway with preservation of adequate laryngeal function for airway protection, swallowing, and voice production. Most recently Wyatt and Hartley (2005) consider laryngotracheal reconstruction (LTR) as an effective surgical procedure, whereas Alvarez-Neri et al. (2005) mean cricotracheal resection (CTR) to be the right way of solution in the management of severe subglottic stenosis. Both authors operated 21 and 22 patients, respectively. *The choice of the operative method, individual judgment, and one's own experiences are essential.*

Undoubtedly, that the outcome of laryngotracheal surgery depends on the grade of the stenosis, the age of the patient, the procedure performed, and the experience of the surgeon influence the result. Most authors report a success rate of 80 to 90% when the patient has undergone a careful preoperative evaluation and when appropriate surgery has been performed. Surgical complications are rare but may occur because of inappropriate stent, graft, inadequate endoscopy, poor follow-up, anterior suprastomal collapse, interactive progression of GERD, or keloid formation (Choi & Zalzal, 1999; Zalzal, Choi, & Patel, 1997).

KEY POINT

Subglottic stenosis may be congenital or acquired. The congenital form is a common laryngeal anomaly due to the defective development of the cricoid cartilage. A typical, hollow inspiratory stridor appears first, as a rule, 2 to 3 weeks after birth when the infant begins to move more. It may also be typical that a catarrhal upper airway inflammation manifests the symptoms. Subglottic stenosis is more often an acquired alteration as a consequence of prolonged intubation. Diagnosis can be established only by means of endoscopy. Its therapy is determined by the measure of the obstruction. The two main surgical methods are incision technique with laryngotracheal reconstruction and cricotracheal resection.

Subglottic Hemangioma

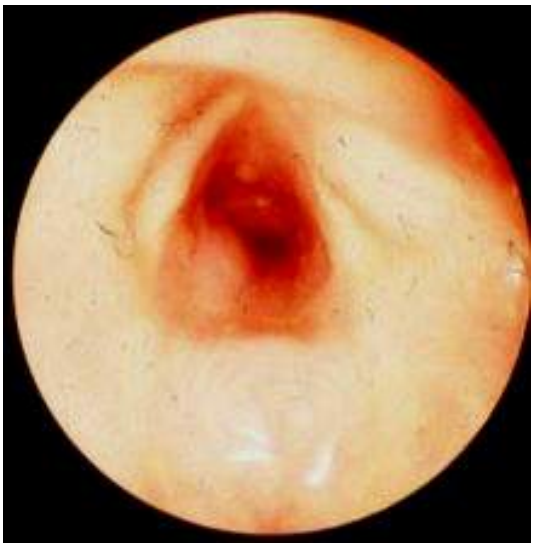
Among the laryngeal neoplasms which may cause respiratory obstruction, subglottic hemangioma, recurrent respiratory papillomatosis and various laryngeal cysts are the most frequently discussed anomalies in the literature. Though hemangiomas often are listed as neoplasms, they are actually manifestations of abnormal blood vessel growth (Myer & Cotton,

1988) or they can be regarded as a hamartoma of blood vessel development (Cotton & Reilly, 1983). Generally, this tumor begins to grow within the first weeks to months of life and may be associated with other vascular birthmarks.

The hemangiomas of the larynx may occur supraglottic, but very seldom; the typical localization is the subglottic region. The lesion is located characteristically in the posterior portion of the subglottis and on the left side (Figures 7-86A and B).

The signs of the subglottic hemangioma manifest themselves first at the age of 1 to 3 months, despite the fact that it is a congenital alteration. The symptoms correspond with those of a pseudocroup: hollow inspiratory stridor, barking cough, clear cry, and in severe forms dyspnea. Derkay and Darrow (2000) observed biphasic stridor, occasionally associated with cough, cyanosis, and hoarseness. *The differential diagnosis between hemangioma and pseudocroup could be rendered difficult, if the symptoms declare themselves in connection with feverish catarrh, but the age helps us in the right diagnosis* in these cases, too. *Laryngitis subglottica does not occur in the first 6 months of life or only very rarely, whereas the symptoms of the hemangioma always appear in this period of life; and if there is no infection, the child is afebrile in contrast to the case of laryngitis subglottica.*

Lateral radiographs may show posterior subglottic masses, but *the anomaly should be diagnosed definitively with endoscopy*. Directly under the vocal cords with intact, sometimes reddish-livid mucosa covered swelling can be seen, at the back and mostly on the left side of the subglottis. Gentle palpation confirms the lesion's softness. An objective diagnosis can be made with histology; a probe excision is, however, contraindicated because of a possible hemorrhage. In the case of a laryngeal hemangioma, also on other parts of the body, hemangiomas or telean-



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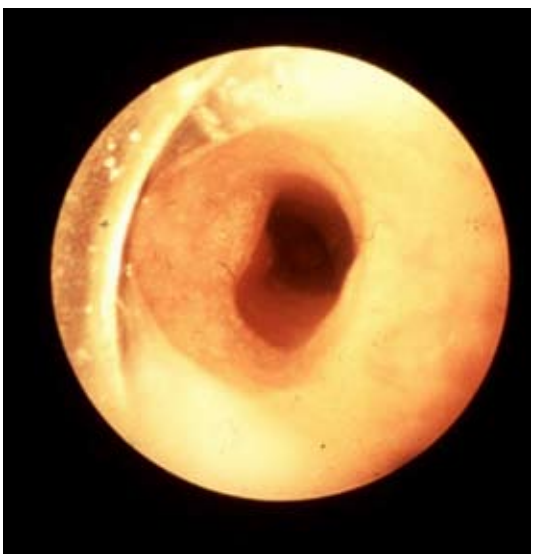


D

FIGURE 7-86. Subglottic hemangioma in a child aged 12 (A) and 14 (B) months, respectively. See the swelling typically located on the back and on the lateral wall of the subglottis.

giectasia may occur in about half of the patients: this observation facilitates the diagnosis. In female infants hemanigoma occurs twice as often as in boys.

The aim of the therapy is to restore normal respiration, attempting to preserve the child's voice and alter the quality of life of both the infant and the family as little as possible (Re, Forte, Berardi, & Mallardi, 2003). Regarding the management, Bent (2006) made with appropriate remarks in his publication: *"A myriad of medical and surgical treatment options exist for children with airway hemangiomas."* The subglottic hemangioma, just like the same alterations on the skin, is susceptible to spontaneous regression; generally it goes back to the age of 3 years. Garfinkle and Handler (1980) summarized with good reason their standpoint two and a half decades ago as follows: "In general, treatment should remain conservative except when there is mechanical obstruction of the airway, hemorrhage, infection, tissue loss, or threatened cardiovascular decompensation." This fact, the spontaneous regression, today also influences the management.

Rahbar et al. (2004) classify the *therapeutical possibilities* under six categories:

1. observation,
2. corticosteroid,
3. tracheotomy,
4. laser,
5. interferon, and
6. laryngotracheoplasty.

We agree with them in that all techniques, above, involve a certain degree of possible effectiveness but no single variant of treatments has been accepted as ideal: all approaches also have their disadvantages and some of them are associated with complications, which can be very serious (Pransky & Canto, 2004). Lasers may be applied conservatively, for isolated lesions but not for circumferential ones because of a possible complication of subglottic stenosis. CO_2 laser was superior to the ND: YAG laser according to the study of Nicolai, Fischer, Truestedt, Reiter, and Grantzow (2005). Re, Forte, Berardi, and Mallardi (2003) prefer repeated endoscopic CO_2 laser treatment of the lesion and perioperative administration of oral cortisone. Patients treated with steroids, however, had recurrence of symptoms when steroid dosage was decreased (Garcia-Casillas, Matute, Cedra, & Vazquez, 2004). Interferon, used in resistant cases, is thought to interfere with endothelial locomotion and, according to Froehlich, Seid, and Morgon (1996) is not utilized because a rapid effect is not expected on the respiratory distress. Naiman, Ayari, and Froehlich (2003) performed open surgical excision in 13 pediatric patients; the cricoid cartilage was left open at the end of the procedure and a postoperative intubation was carried out. Extubation after surgery was successful in all cases. The risk of a postoperative subglottic stenosis was limited. This strategy enabled tracheotomy to be avoided in all but 7.2% of all cases. Open surgical excision of subglottic hemangiomas can be performed as a single procedure, avoiding a tracheostomy, when modern surgical techniques developed for laryngotracheal reconstruction are incorporated (Vijayasekaran, White, Hartley, Rutter, Elluru, & Cotton, 2006).

In other, mostly earlier, publications one can read also about other therapeuti-

cal methods such as roentgen irradiation (Ferguson & Flake, 1961), sclerotization (Pierce, 1962), cryocauterisation (Jokinen, Palva, & Kärjä, 1981), and stiching with radioactive gold (Holborow & Mott, 1973), but these interventions are not in practice anymore.

The management is influenced by the degree of the respiratory obstruction. After evaluating the success and complication of variable treatment options of congenital subglottic hemangioma in 372 patients, the conclusion of Bitar, Moukarbel, and Zalzal (2005) was the following:

- treatment should be individualized;
- the CO2 laser is useful when used cautiously;
- steroid may be beneficial;
- excision is for stubborn cases.

According to Benson et al. (2006), as hemangiomas usually regress by the age of 12 to 18 months, any surgical or medical interventions should be based on the severity of symptoms. The Budapest team (Lellei, 2004; Lellei, Farkas, & Hirschberg, 1978; Lellei & Hirschberg, 1989) represents in mild cases the wait-and-see conception, in severe cases tracheotomy is performed by us, and generally by the age of 3 to 4 years after suitable spontaneous regression of the symptoms decannulation is possible. Neither in our patient data, nor in the cases reported in the literature, has return of the hemangioma been observed after regression in childhood. Disadvantage of tracheostomy may be speech developmental delay (Nicolai, Fischer-Truestedt, Reiter, & Grantzow, 2005). In our opinion, whichever surgical procedure is applied, must be made individually and very cautiously to prevent any damage to the structures in the larynx (voice) or the subglottic area.

KEY POINT

The typical symptoms of a subglottic stenosis (clear cry, hollow inspiratory or biphasic stridor, barking cough) are the same in various distinct instances. Accordingly, the identification of a correct diagnosis may be complicated. The history may help: laryngitis subglottica (pseudocroup) occurs in the age of half a year at earliest, always with fever. The congenital rigid stenosis and also the congenital hemangioma manifest themselves in the first 6 months of life, both are afebrile. Hemangioma in the larynx is almost always subglottic and is mostly located on the back and left side of this region. There are several treatment possibilities. As a spontaneous regression can be expected at an age of 3 to 4 years, many authors accept the wait-and-see standpoint; in cases with severe symptoms tracheotomy should be performed; after regression of the tumor the decannulation is always possible. Other management versions are: steroid, interferon, laser, open surgery.

Contusion (injury) of the Larynx

Being elastic, the larynx of infants and young children is more resistant to trauma than that of adults. Thus, bunt laryngeal trauma is frequently overlooked in children (Myer, Orobello, Cotton, & Bratcher, 1987). Fracture, dislocation of the cartilages, bleeding, or edema of the mucous membranes nevertheless may occur.

The nature of acoustic signs largely depends on the site and type of trauma. The rapid insult may cause laryngospasm like a foreign body aspiration with stridor, short apnea, and aphonia (Figure 7-87).

Suffusion and uneveness of the margin

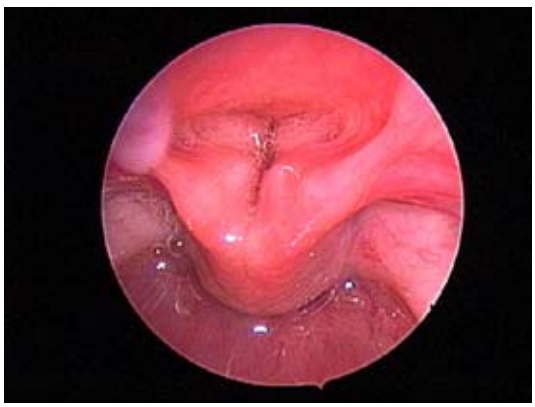


FIGURE 7-87. Laryngospasm in a child.

of the vocal cords make the voice veiled or hoarse. Whenever differences between the levels of the two sides of the larynx are present, structural changes of the voice and a compensatory pseudovocal cord voice may be found. If the lumen is narrowed, stridor-phonation develops. *When the trauma leads to the development of a subglottic hematoma the symptoms will be similar to those of pseudocroup; however, without fever and catarrh, that is, a clear or dull cry, a barking or hollow cough, and an inspiratory stridor of subglottic character appear* (Figure 7-88).

KEY POINT

The larynx of infants and young children is more resistant to trauma than that of adults. The possible presenting acoustic signs depend on the site of trauma. Rapid insult may cause laryngospasm with short apnea and aphonia, wheras suffusion of the vocal cords makes the cry or voice veiled or hoarse. In the cases with subglottic lesion the typical sound phenomena characteristic of this region develop.

Diagnosis should be ascertained by flexible or rigid laryngoscopy. In mild cases observation seems to be enough; in severe cases with serious breathing difficulty tracheostomy and early surgical intervention appears to be the treatment of choice (Kurien & Zachariah, 1999).

Tracheal Changes

The localization of tracheal changes can be intratracheal, extratracheal, and intramural.

Congenital Goiter

This is a relatively rare form of tracheal stenosis due to an extratracheal change. *The congenital goiter compresses the upper portion of the median segment of the trachea anteriorly and laterally.*

The symptoms (stridor, coughing, occasionally dyspnoea) are present from birth, diminish on the posterior tilting of the head (extension of the trachea), and may subside within a few days.

The sound phenomena produced may resemble those heard in tracheal stenosis; in a patient of ours a deep, hollow cough was noted (Figure 7-89).

The diagnosis does not cause any problem.

The treatment is to give thyroid hormone in cases where the spontaneous regression comes late.

KEY POINT

Congenital goiter is a rare anomaly but its knowledge is advantageous in order to differentiate it from other tracheal obstructions. Stridor, coughing, occasionally dyspnea are present from birth. The symptoms are the consequence of compression of the upper portion of the trachea, possibly both anteriorly and laterally. The diagnosis is always unambiguous.

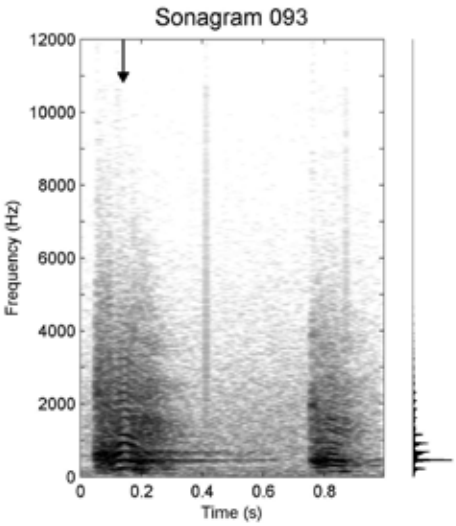


FIGURE 7-88. Contusion of the larynx, subglottic haematoma. Child aged two and a half years. *History:* Fell on the handle bars of a bicycle; difficult breathing and suffocating coughs ever since. *Laryngoscopy:* Grayish-blue bulges on both sides directly underneath the vocal cords. *Acoustic finding:* Hollow cough, occasionally stridor. *Sonagram of the cough No. 93:* Two separate cough pockets in time between 0.05 to 0.35 s, and 0.75 to 0.9 s. The first cough seems to be more definite than the second one. Marked, intensive noise components up to 2800 Hz. At 3400, 4100, and 5500 Hz thin ribs about 250 Hz apart appear making the cough hollow. It is low-pitched due to the absence of high-frequency noise components. The amplitude cross-section is of particular value in the acoustical evaluation of this case because of the dense noise band.

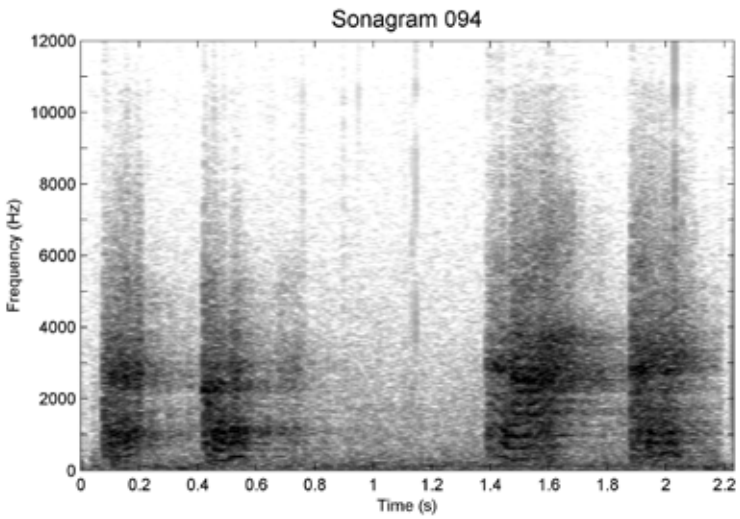


FIGURE 7-89. Congenital goiter. Infant aged 2 weeks. *History:* Admission on the day of birth with asphyxia, dyspnea, perioral cyanosis. The enlarged thyroid, the size of a green nut, is palpable on both sides of the neck. *Laryngoscopy:* Intact, mobile vocal cords, subglottic space free. *Acoustic finding:* Deep, hollow cough. When crying or when handled, stridor with cyanosis. *Sonagram No. 94:* Noise bands showing different arrangements. The amorphous band between 2500 and 10000 Hz can be considered dominant. The noise over 6500 Hz makes the cough dull, whereas its hollowness is due to the more regularly arranged noise blocks mimicking a dense harmonic structure between 0.05 to 0.25 s, in the range 0.45 and 0.6 s and further between 1.4 to 1.75 s and 1.85 to 2.25 s. A low fundamental vibration is responsible for the low pitch of the cough. This can be seen in the record as ribs of noise arranged at 180 to 200 Hz distance between 1.5 and 1.8 s.

Vascular Anomalies

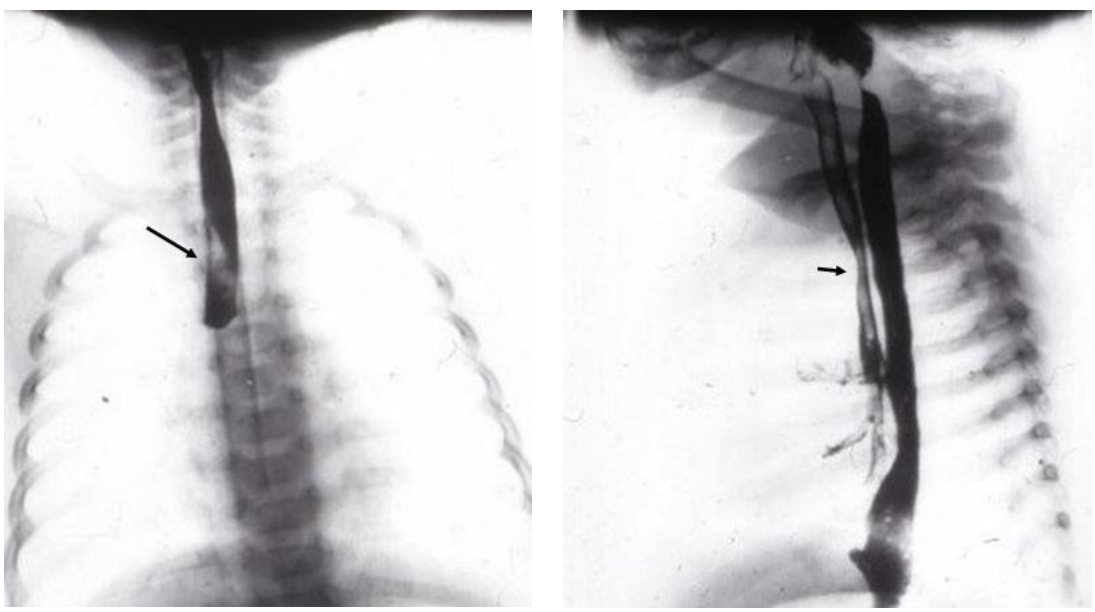
The five *most common forms of congenital vascular anomalies* causing compression of the trachea are:

1. double aortic arch,
2. right aortic arch with left ligamentum arteriosum which also makes a vascular ring,
3. aberrant right subclavian artery (arteria lusoria),
4. anomalous innominate artery (brachiocephalic trunk),
5. anomalous left common carotid artery or pulmonary artery.

The most common among them is compression from the innominate artery, and the second one is the complete vascular ring, also known as the double aortic arch (Bove et al., 2001; Eklöf et al., 1971; Fearon & Shortreed, 1963; Nadas & Fyler, 1972).

The aberrant blood vessels exert pressure on the trachea anteriorly (Figures 7-90A and B) *or posteriorly* (if the blood vessel is located between the trachea and esophagus). *The vascular ring, on the other hand, encircles the trachea and esophagus*, *compressing both* (Figures 7-91 and 7-92A, B, C, and D). Absence of esophageal compression, thus, does not rule out vascular anomaly. The trachea, itself, is inherently normal.

Depending on the severity of tracheal compression, the symptoms present themselves immediately after delivery or during the first year of life. Attacks of apnea, cyanosis, tachypnea, recurrent respiratory infections, dysphagia, hyperextended posture of the infant's neck (in an effort to straighten the compressed airway), occasionally reflex



A B

FIGURE 7-90. Tracheogram (A, A–P) and tracheoesophagogram (B, lateral) of a stenosis tracheae due the impression of truncus brachiocephalicus. Two-and-a-half-month-old infant.

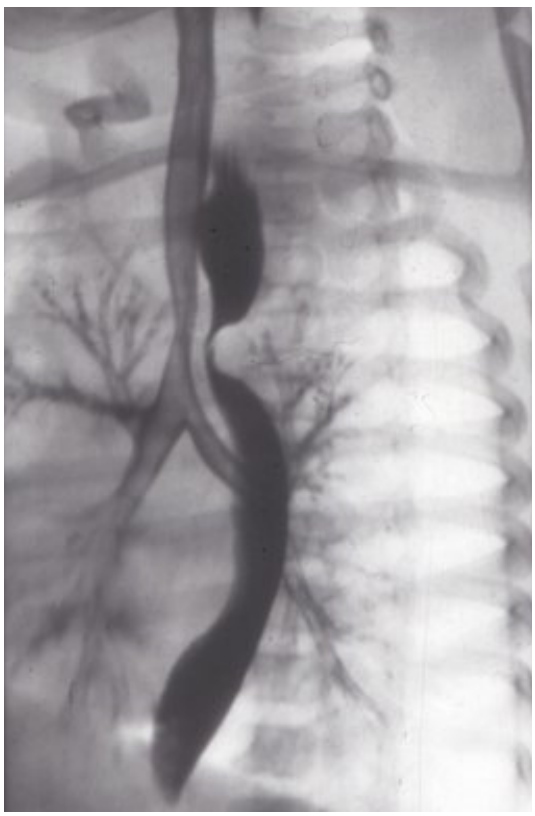


FIGURE 7-91. Tracheoesophagogram showing the stenosis of both the trachea and esophagus due to aorta duplex in a child age 10 months. See the circular impression.

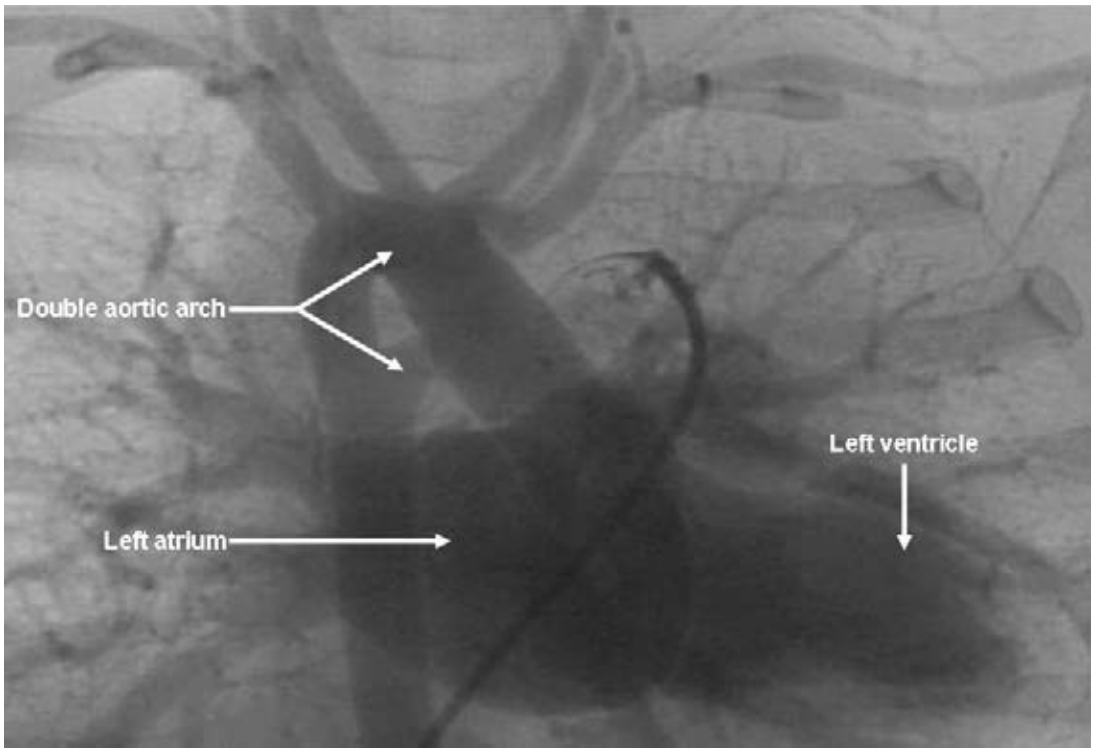
KEY POINT

The most common vascular anomalies causing compression in the trachea are the double aortic arch and the innominate artery (truncus brachiocephalicus). The vascular ring encircles the trachea and the esophagus, the arteria anonyma exerts pressure on the trachea anteriorly. Depending on the severity of the tracheal compression, dyspnea, cyanosis, recurrent respiratory infections, dysphagia may be present. Among the pathologic sound phenomena the noisy, continuous stridor is characteristic which may occur in both respiratory phases. Hollow coughing sound also may be observed, but the cry is always clear. Endoscopy, contrast x-ray methods, and MRI verify the diagnosis. The surgical indication belongs to the competency of the cardiac surgeon; however, a close collaboration with the otolaryngologist and bronchologist is essential.

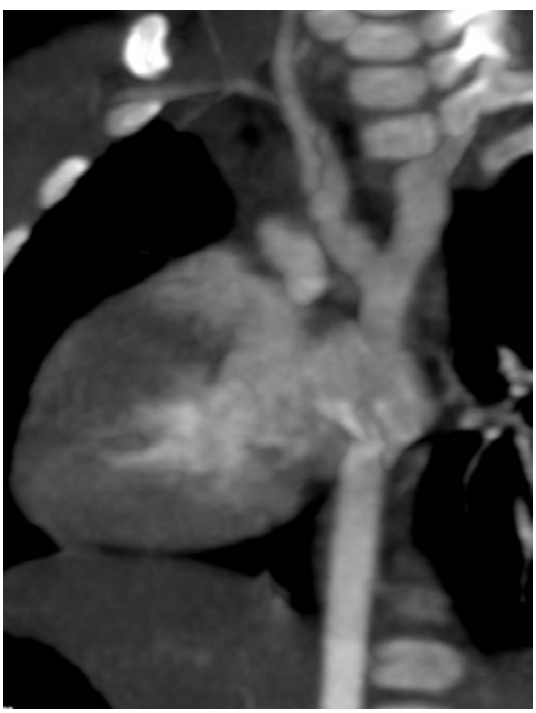
apnea, and most frequently pathologic sound phenomena indicating respiratory disorder and airway obstruction are the characteristic symptoms.

The acoustic findings include biphasic, inspiratory or, infrequently, expiratory stridor of tracheal character (which is noiselike, not phonationlike; continuous, and not interrupted), or a deep or hollow stridor; a hollow coughing sound (which may also be ringing according to the data in the literature); and a clear cry. A transition from phonation into coughing may also occur. A considerably prolonged expiratory phase also may exist.

In typical cases of a vascular ring causing severe obstruction, a marked stridor tends to dominate the picture (Figure 7-93), whereas if there is only tracheal impression due to an anomalous innominate artery, coughing is mainly observed. The symptoms and the characteristic sounds may elicit the suspicion of vascular anomaly. Bronchoscopy reveals the pulsatil vascular impression on the trachea, usually on the anterior wall, 12 cm above the carina (Figure 7-94). Esophagoscopy may detect extrinsic esophageal compression. The barium swallow study demonstrates the circumferential narrowing of the trachea. Further clarification of the anomaly and differential diagnosis are facilitated by angiography (Berdon, Baker, Bordiuk, & Mellins; 1969, Eklöf et al., 1971), echocardiography and color-flow Doppler,



A



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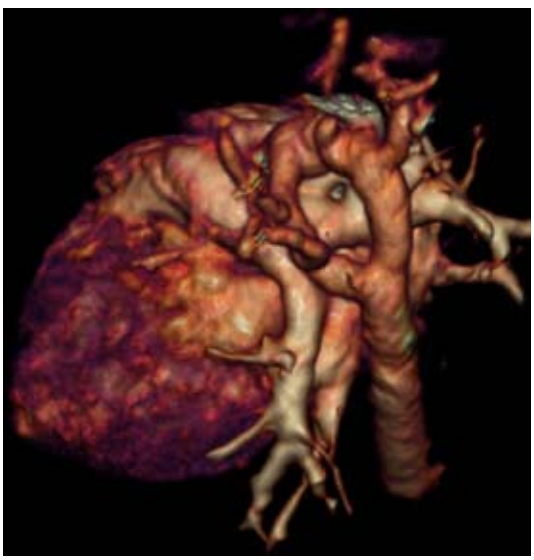
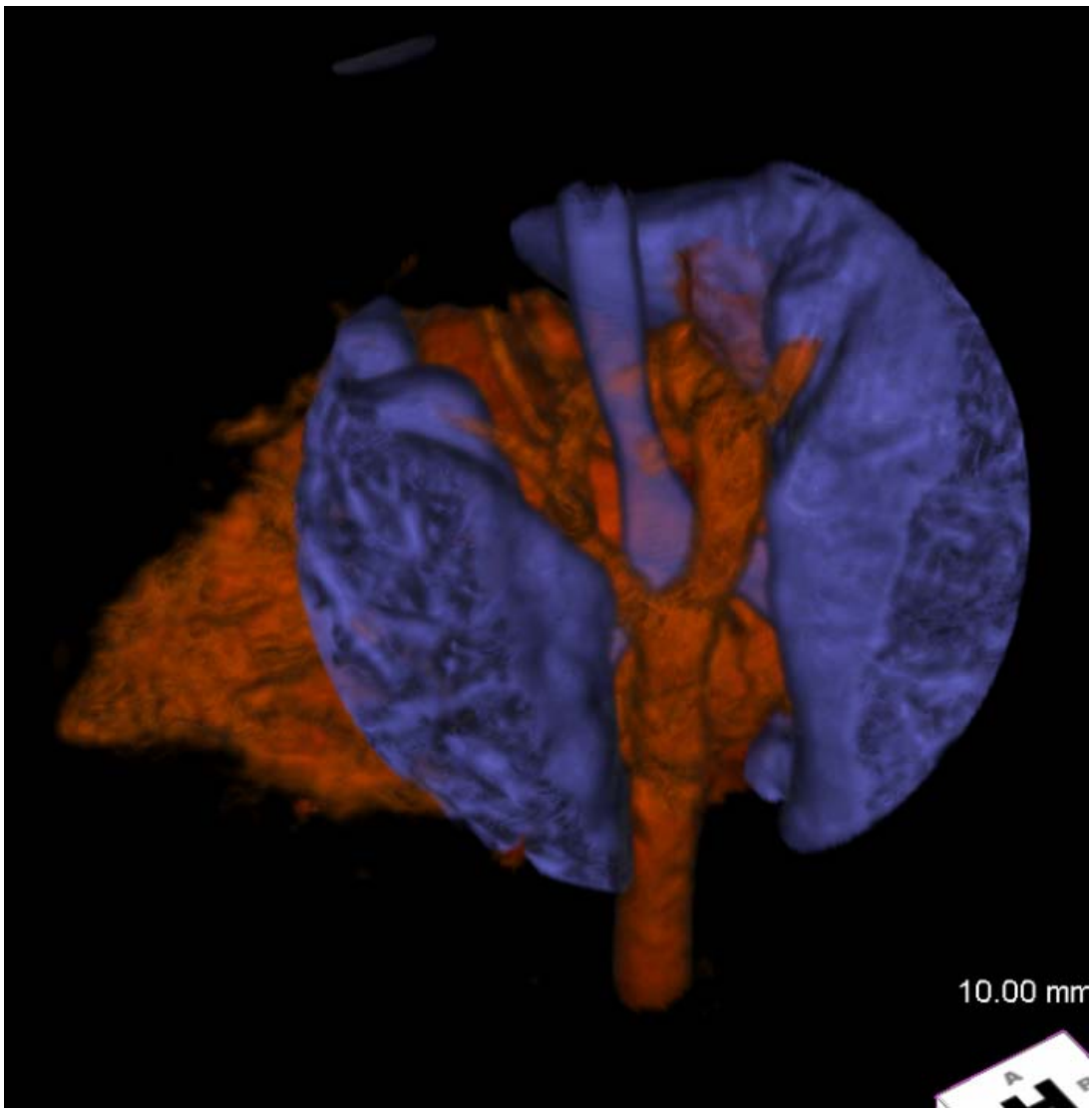


C

FIGURE 7-92. A. Angiogram shows an aorta duplex in a 1-month-old infant. **B**, **C**, and **D**: Pictures from www.ctcardiac.com : Double aortic arch encircling and compressing the trachea in a 2-month-old infant.



D

computerized tomography scan, magnetic resonance imaging, digital subtraction angiography, aortic angiography, and cardiac catheterization (Bhimiji & Eggerstedt, 2007). Simultaneous tracheoesophagography (Csermely et al., 1974) and tracheoangiography (Hirschberg & Lellei, 1982) gave significant help some decades ago in the diagnosis, to determine the relation between the trachea and esophagus and the blood vessels as well. These useful methods, however, have gradually lost significance. Magnetic resonance imaging has become the standard means of imaging pediatric airway obstruction due to vascular anomalies; however MRI requires a long acquisition time and is prone to motion artifacts. The development of spi-

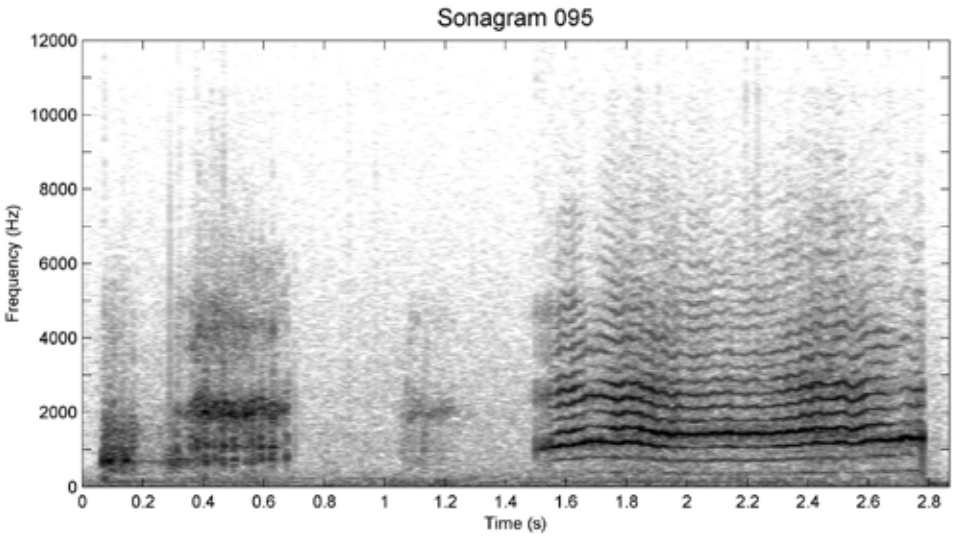


Figure 7-93. Congenital vascular anomaly, double aortic arch (corrected surgically later on). Infant aged 10 months. *History:* From 4 months of age, after having started to eat with a spoon, the infant's mother noticed stertorous breathing and cough. Treated repeatedly for wheezy bronchitis. *Laryngobronchoscopy:* Thick, mobile vocal cords; indistinct bulge in the trachea. *Acoustic finding:* Strong, continuous, inspiratory stridor of tracheal origin. *Sonagram No. 95:* 0.3 to 0.6 s: Marked noise components around 2000 Hz and between 4000 and 5000 Hz. Regular harmonic structure in the 1.5 to 2.8 s time range, clear from noise up to 10000 Hz. No melody structure.

ral or helical computed tomography provides an alternative imaging modality for evaluating pediatric airway obstruction (Gustafson, Liu, Link, Strife, & Cotton, 2000) The standard diagnostic method is still, however, endoscopy (Bailey & Calhoun, 1993; Berrocal, Madrid, & Novo, 2004). Virtual bronchoscopy (VB) is a novel technique for noninvasive evaluation of the tracheobronchial tree (Jones & Athanasiou, 2005). We agree, however, with De Wever et al. (2004) in that it will never replace actual bronchoscopy as it does not currently enable the detection of subtle mucosal lesions and is not sensitive enough to detect dynamic movement problems which may cause airway obstruction (Burke et al., 2000).

Diagnosis, management and postoperative care of infants and children with tracheastenosis due to vascular anomaly need a very close collaboration among pediatricians, cardiologists, cardiac surgeons, radiologists and laryngobronchologists (Lozsádi et al., 1981). The surgical indication is the competency of the (pediatric) cardiac surgeon. Surgical correction involves suspending the innominate artery or the aorta anteriorly to the sternum (i.e. inominopexy or aortopexy), and in cases of aorta duplex the treatment consists of surgically dividing the ring. In pediatric patients undergoing surgical repair of congenital cardiac defects are predisposed to laryngeal anomalies owing to (1) frequent intubation, (2) prolonged ventilatory support, (3) recurrent laryngeal nerve injury, and (4) subglottic stenosis (Khariwala, Lee, & Koltai, 2005). After cardiac surgery, laryngobronchoscopic control is suggested, especially in patients, where a secondary tracheal cartilage deformity (tracheomalacia) could presently evolve by the resulting pressure. The weakness

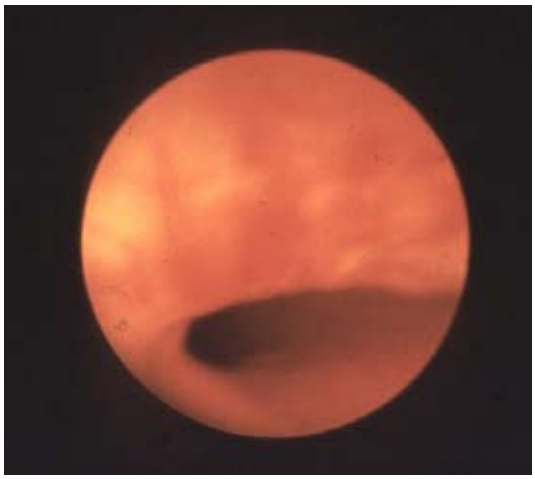


FIGURE 7-94. Endophoto in the same infant (see Figure 7-91): tracheal obstruction due to vascular anomaly.

of tracheal cartilages presenting with stridor may even worsen immediately following vascular repair, because the external supporting structure has been removed.

Paratracheal or Parabronchial Lymphadenitis (thoracic lymph node, enlargement, lymphadenopathy, bronchial rupture)

Lymphadenopathy used to be concomitant with tuberculosis or also occur as an independent tuberculotic entity. Because of the possibility of suffocation due to bronchial rupture, it was a dreaded complication of tuberculosis (Görgényi-Göttche, 1958).

Tuberculous affection of the regional lymph nodes of the lower respiratory tract rarely occurs today, whereas a nonspecific enlargement of the glands (Kallay, Csermely, & Hirschberg, 1966), occasionally with invasion of the bronchi, some decades ago, was not so uncommon. Recently, Andronikou et al. (2004) gave an account of tuberculous mediastinal lymphadenopathy in children from the Department of Paediatric Radiology, Red Cross Children's Hospital, Cape Town, South Africa: lymph nodes were present in 92 patients among 100 children with clinically suspected primary pulmonary tuberculosis. The most common location was the subcarinal position. Bronchial compression was identified in 29 patients, most commonly, the left main bronchus was involved. According to the authors plain radiographs may be unreliable and CT is the current "gold standard" for demonstrating this.

Apart from the general symptoms of slow onset and low fever in specific cases, sudden onset and high fever in nonspecific cases, the symptoms are identical in both forms and are due to compression or obstruction of the lower portion of the trachea and/or of the bronchi.

A typical cough may be observed in paratracheal lymphadenopathy which has a typically brassy ring (Figure 7-95). When a caseous or purulent lymph node ruptures and invades the bronchi, coughing increases and there is dyspnoea the severity of which depends on the degree of obstruction.

KEY POINT

The lymph nodes compressing the trachea and/or the bronchi cause typical brassy ringing cough, on the basis of which our ancestors diagnosed the "bronchial rupture" of tuberculotic lymph nodes. Today a nonspecific enlargement of the glands, occasionally with invasion of the bronchi, may develop.

The diagnosis can be made by radiologic procedures especially by CT and by

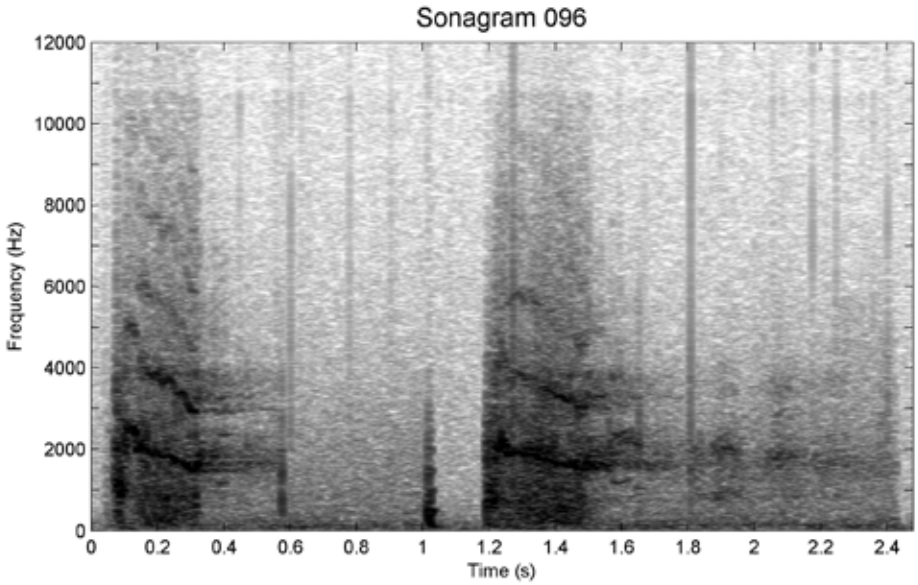


FIGURE 7-95. Parabronchial (nonspecific) lymphadenitis. Infant aged 3 months. *History:* Cough and fever for 2 to 3 weeks. *Bronchoscopy:* Circular narrowing of the orifice of the right main bronchus mainly laterally. *Tomography:* Most likely, plaque of lymph nodes in the bifurcation area. *Acoustic finding:* Brassy, ringing cough. *Sonagram No. 96:* Very well structured noise with marked overtonelike components arranged at considerable intervals and containing large glides (approx. in 1000 to 1500-Hz steps).

MRI; bronchoscopy reveals the tracheal/bronchial compression.

The management concerns to the pulmonologist and depends on the etiology.

Mediastinal Tumors

Several kinds of mediastinal tumors are known which differ as regards their localization and morphology. Tumors of the thymus and teratoids develop in the anterior mediastinum (exceptionally a hyperplastic but not tumorous thymus also may narrow the tracheal lumen if the tracheal wall is soft), enterogenic cysts (Figure 7-96) tend to occur in the middle, whereas neurogenic tumors are more common in the posterior mediastinum. Tansel et al. (2006) treated 187 infants and children with primary mediastinal mass between

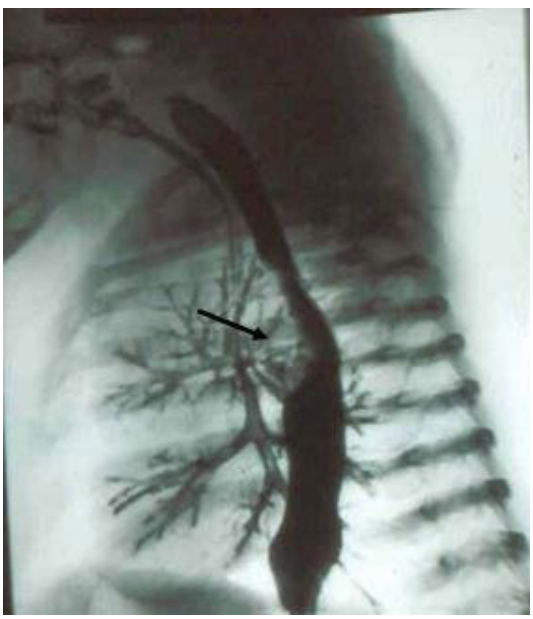


FIGURE 7-96. Bronchography about a mediastinal cyst in a 2-year-old child.

1980 and 2004 at the Department of Cardiovascular Surgery, Istanbul University. The cases were in the order of frequency: lyphoma, neurogenic tumors, cystic lesions, and cardiac tumors.

In the cases of mediastinal tumors pathologic sound phenomena are due either to the compression of the trachea or to recurrent paresis arising as a result of pressure exerted on the left recurrent nerve in the mediastinum. The symptoms of unilateral recurrent nerve paresis (stridor-phonation, pathologic cry) were described previously in this chapter. Compression of the trachea leads to attacks of suffocation, stridor, and intractable coughing (Kallay & Hirschberg, 1963). *The stridor may be tracheal and deep hollow in character, whereas the cough is hollow or ringing. In general, a ringing cough indicates a more distal localization of the tumor*. In our case a brassy, ringing cough was heard (Figure 7-97). *When there is no recurrent nerve paresis, the cry is clear and regular.*

KEY POINT

Several types of tumors may develop in the mediastinum: thymoma in the anterior part, enterogenic cysts in the middle, and neurogenic tumors in the posterior part of the mediastinum. The specific sound phenomena to be heard come about due to tracheal compression or due to the left recurrent nerve paralysis, if the latter is involved in the pathological process. Compression of the trachea may cause suffocation, deep hollow stridor, and ringing cough. Stridor and cough may be the symptoms of recurrent nerve paralysis as well.

Diagnostic evaluation includes: chest x-ray, CT scan, MRI of the chest, CT-

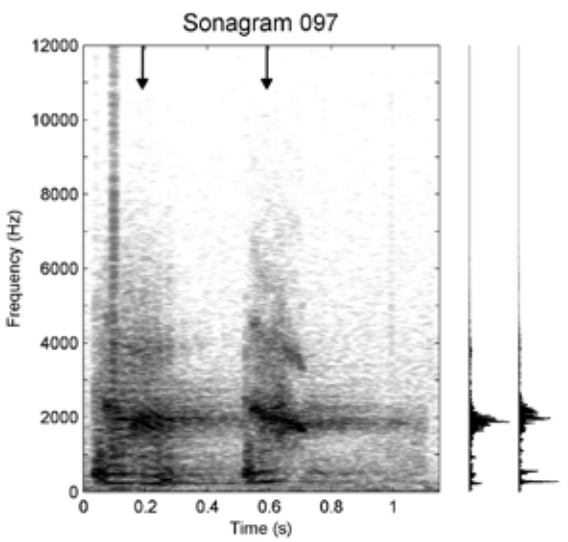


Figure 7-97. Mediastinal tumor (sarcoma). Child aged three and a half years. *History:* Worsening cough for a few weeks; no fever. *Chest x-ray:* Strongly enlarged cardiac shadow, obscuring the trachea. *Laryngoscopy:* No laryngeal change. *Bronchoscopy:* Not performed. *Acoustic finding:* Cough, in other instances stridor with dyspnea. *Sonagram No. 97:* Noise components throughout the entire width of the spectrum. Around 1800, 2100, 4000, 5000, 6000, and 6500 Hz they are thickened and show a scanty overtonelike arrangement lending the cough a metallic ring. In the full frequency range a thin noise component in the 0.09 to 1.1-s time-scale range

guided needle biopsy, mediastinoscopy with biopsy, and bronchoscopy (to reveal possible airway obstruction). The laryngobronchologist may help the other specialists in the diagnosis and occasionally in the management as well. Treatment varies based on the type of tumor.

Tracheomalacia (functional stenosis of the trachea, soft trachea)

Congenital tracheomalacia (TM) resembles laryngomalacia and the two anomalies may be associated. *This term, like laryngomalacia, has been adopted in the literature as a conceptual category and is misleading because it is not a sui generis malacia, that is, a secondary destruction of cartilage, but a congenital softening and flatness of the tracheal cartilage* (Händel & Wunderlich, 1968). Information on TM could already be obtained in the 1960s and 1970s from the publications of Ruhrmann (1963, 1964), Picot et al. (1969), Holinger, Zimmerman, and Schild (1972), and Minnigerode (1972), but this malformation of the trachea is also discussed rich in detail in several up-to-date pediatric otolaryngologic textbooks (Bluestone et al., 1996; Cotton & Myer, 1999).

According to Carden, Boiselle, Waltz, and Ernst (2005) malacia refers to "softness" and, in medical terminology, generally refers to cartilage or bone. TM refers to a weakness of the trachea, frequently due to reduction and/or atrophy of the longitudinal elastic fibers of the pars membranacea, or impaired cartilage integrity, such that the airway is softer and more susceptible to collapse. The normal intrathoracic trachea dilates to some extent with inspiration and narrows with expiration. In TM, there is an accentuation of this physiologic process: an excessive narrowing is prominent during forced expiration, cough or the Valsalva maneuver. Thus, TM is characterized by an expiratory tracheal collapse (Bove et al., 2001; Sudre-Levillain, Roman, Nicollas, & Triglia, 2001). The deformity of the trachea is particularly conspicuous on expiration when, under the effect of extrabronchial intrathoracic pressure, the flattened anterior wall almost touches the bulging pars membranacea (Figures 7-98A, B, and C).

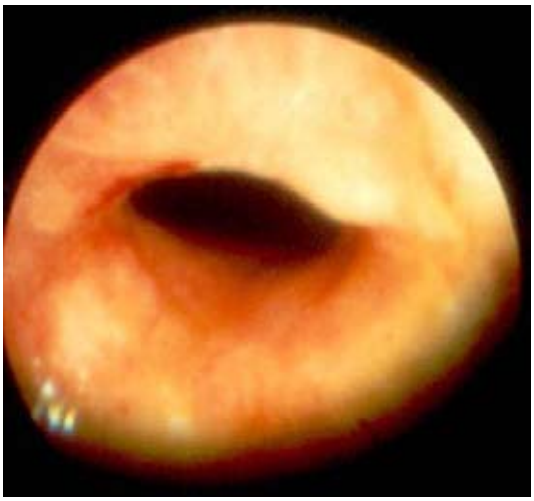
Mostly the middle and lower thirds of the trachea are affected, or may involve the entire trachea. If the mainstem bronchi are involved as well, the term tracheobronchomalacia is employed.

As the stenosis is not rigid, it is easily passable by the bronchoscope; moreover, the walls of the trachea can be parted even by means of O_2 overpressure during bronchoscopic examination in relaxation. This is the reason why its traditional labeling is functional stenosis.

Primary airway malacia is not rare in the general population with an estimated incidence of at least 1 in 2, 100 children (Boogard, Huijsman, Pijnenburg, Tiddens, Jongste, & Merkus, 2005). Among our patients, observed during 6 years in Heim Pál Hospital for Sick Children, Budapest (Table 7-1), tracheomalacia was the most freqent cause of tracheal stenosis (Hirschberg, 1989; Hirschberg & Lellei, 1982). However, it may be difficult to recognize it on the basis of clinical features that overlap with those of more common pulmonary diseases such as recurrent lower airway infection, therapy resistant, or atypical asthma.

In its classical form, the signs and symptoms of TM occur not at birt but during the

first weeks to months of life. Dyspnoe, and episodes of cyanosis may call attention to the disease, but the most characteristic symptoms are auditory, indicating unequivocally the tracheal localization of the change. Stridor, coughing, and occasionally pathologic cry happen to appear, although the three are not necessarily concomitant. *All three are typically hollow in character* (Figure 7-99). According to Sudre-Levillain et al. (2001) the revealing clinical features (stridor, wheezing, chronic cough, apneic attack, and difficult breathing) usually occur after a symptom-free period during the first year of life (during the first 3 months in 60% of the cases). *The stridor may be inspiratory, biphasic, or expiratory. If there is malacia of the extrathoracic trachea, inspiratory stridor can be found. The expiratory stridor resembles wheezing* and the cough (which is marked when there is an associated catarrh, a common complication) resembles the barking of the sea lion (see Figure 6-57). Exceptionally it may also have a brassy ring. The individual sound phenomena may be clustered, for example, a regular, then hollow cry may be interrupted by hollow cough. If tracheomalacia is associated with laryngomalacia, a cackling stridor characteristic of the latter can also be observed. Persistent or recurrent pneumonia belongs to the main symptoms and also atelectasis on the chest radiograph is a common sign (Yalcin, Dogru, Ozcelik, Kiper, Aslan, & Gozacan, 2005). Usually, but not always, the symptoms gradually improve.



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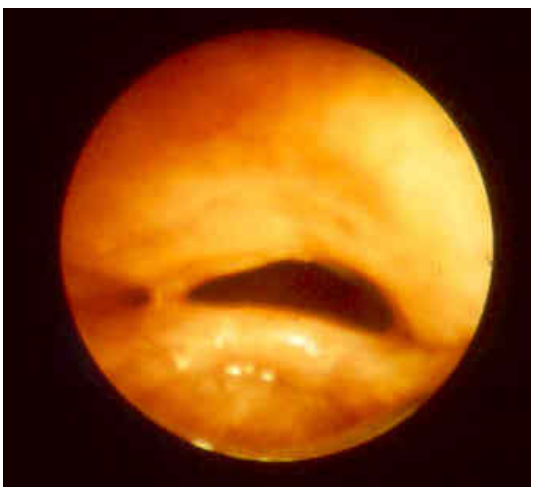

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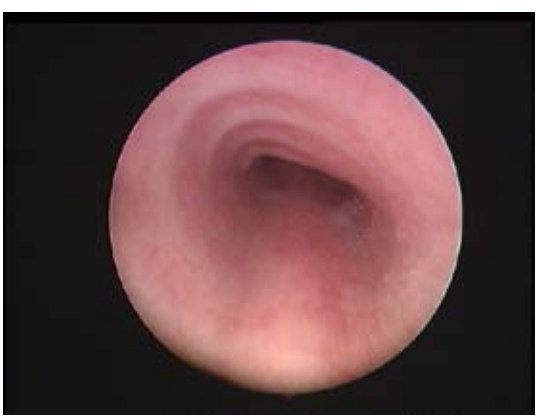

C

FIGURE 7-98. Functional stenosis of the trachea (tracheomalacia) in infants aged 9 (A), 10 (B), and 11 (C) months, respectively. The flattened anterior wall of the trachea almost touches the bulging pars membranacea.

TABLE 7-1
Distribution of Tracheal Stenoses of Infants and Children, Observed in Heim Pál Hospital for Sick Children, Budapest During Six Years (1981–1986)

Diagnosis	Age				
	0-1 years	1-3 years	3-6 years	6-14 years	Altogether
Struma	1				1
Thymus	1				1
Vascular anomaly	13	3	2	3	21
Mediastinal tumor (sarcoma, cysts)	1		1	1	3
Lymphatic ganglion			1	1	2
Cicatrisation due to prolonged intubation	1		3	6	10
Functional stenosis (tracheomalacia)	91	23	6	1	121
Dyskinesia	4	3	2	2	11
Rigid circular stenosis	2				2
Circumscript anomaly of cartilage	1				1
Tracheostomy, granuloma, fibroma		5			5
Hemangioma		1			1
Exogen foreign body	11	5	2	3	21
Endogen foreign body: crust, fibrin		4		1	5
Altogether	126	44	14	12	205

NOTE: The number of tracheal cicatrization due to intubation or stent has been grown gradually since this study.

KEY POINT

Tracheomalacia is not a sui generis malacia, but a congenital softening and flatness of the tracheal cartilages. The deformity of the trachea is particularly conspicuous on expiration when the flattened anterior wall almost touches the bulging pars membranacea. The stenosis in not rigid, it is easily passable by the bronchoscope. All three sound phenomena, that is, cry, stridor, and cough, that occur are hollow in character. Dyspnea and mild, episodic cyanosis may be present from the first weeks of life. The expiratory stridor resembles wheezing. Diagnosis can be established by endoscopy, contrast x-ray pictures, and CT. The symptoms gradually improve, the need for surgical management is rare.

Diagnosis can be established by endoscopy. We agree with Boogard et al. (2005) in that *bronchoscopy is an inevitable, essential, and mandatory tool and* that *it should remain the preferred method for evaluating the airways*. However, this view is debated in pediatrics because endoscopy is in essence invasive and preliminarely requires that the child can spontaneously breathe. If the child is paralyzed, heavily sedated, airway collapse may arise, but as the anesthesia is reduced and the patient begins to move and cough, the dynamics of the airway can be assessed entirely. The length and position of the stenosis, and expiratory and inspiratory changes in the lumen can be well demonstrated by airway

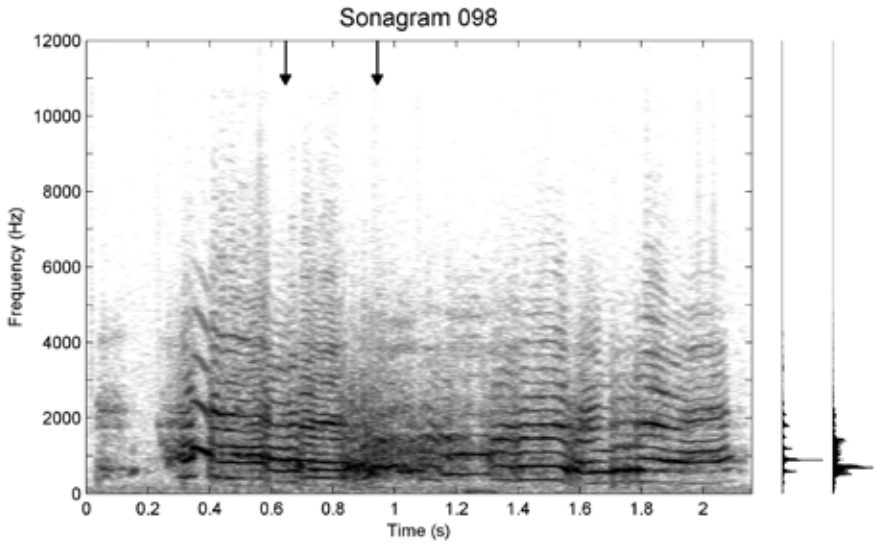

FIGURE 7-99. Functional stenosis of the trachea (tracheomalacia). Infant aged 9 months. *History:* Inspiratory stridor from birth. One week before admission upper respiratory tract catarrh with an attack of apnea. *Laryngoscopy:* No pathologic changes in the larynx. *Tracheoscopy:* The anterior and posterior walls of the trachea bulge forward over a segment of a few cm about the middle of the trachea. This results in a stenosis of approximately 2 to 6 mm diameter, which is passable and not rigid. Contrast esophagogram: normal. *Acoustic finding:* Hollow, coughlike phonation (or stridor). *Sonagram No. 98:* Harmonic structure of variable density. Some interharmonic distances are normal (400-500 Hz), others are short (180-200 Hz) making the sound hollow. These follow one another without transition. Some overtones, for example, the ones around 1000, 2000, and 4500 Hz, are noisy and thickened.

fluorography. The latter procedure may be useful for ruling out the presence of vascular anomalies (Csermely et al., 1974). Using contrast material, the lateral photo demonstrates the (sometimes very severe) narrowing of the trachea, whereas the A-P picture shows normal circumstances (Figure 7-100).

In addition to bronchoscopy and/or -graphy, further information, concerning localization, extent, collapse, stability of the tracheal wall, distal portions of the stenosis, and extraluminal compressions *may be obtained by spiral CT* (Heussel, Hafner, Lill, Schreiber, Thelen, & Kauczor, 2001).

In many children with TM, intervention is not necessary. As the child grows the tracheal cartilage strengthens and

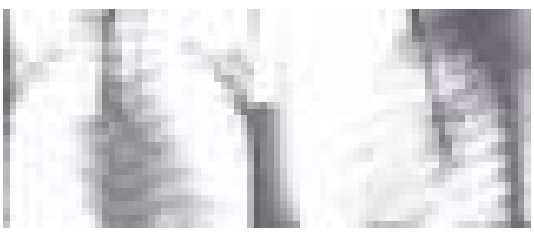

FIGURE 7-100. Tracheography in the same infant with functional stenosis of the trachea (tracheomalacia). The lateral view (right) demonstrates a severe obstruction, the A–P picture (left) shows normal width of the trachea.

stiffens. The symptoms often resolve in children by the age of 1 or 2 years. Therefore, *conservative therapy* (humidified oxygen therapy, pulmonary physiotherapy) *is preferred* (Boogard et al., 2005). Continuous positive airway pressure is an effective treatment for infants with mod-

erate-to-severe tracheobronchomalacia. Surgical treatment is indicated only rarely, in severe and secondary forms (Sudre-Levillain et al., 2001), when the patient is unresponsive to medical management, or when life-threthening symptoms occur. Treatment with tracheopexy, tracheal resection, tracheal reconstruction, and external splinting with autologous materials have been attempted with limited success. During the past decade, aortopexy (Abdel-Rahman, Ahrens, Fieguth, Kitz, Heller, & Moritz, 2002), and aorto-pericardio-sternopexy (Schaarschmidt, Kolberg-Schwerdt, Pietsch, & Bunke, 2002) have become the procedure of choice for management of severe forms but, to date, no perfect operation exists for the treatment of severe tracheomalacia.

Circumscribed Congenital Anomaly of the Tracheal Cartilages (individual cartilage deformity)

The *three most common forms of intramural tracheal stenosis* are:

1. tracheomalacia with or without dyskinesis (over longer segments the cartilages of the trachea are soft and flattened, and also the tone of the posterior wall made up of connective tissue is decreased; see the next section;
2. rigid stenosis (a few tracheal cartilages form a ring, and the membranous part is absent; see upcoming chapter);
3. deformity of one or two tracheal cartilages.

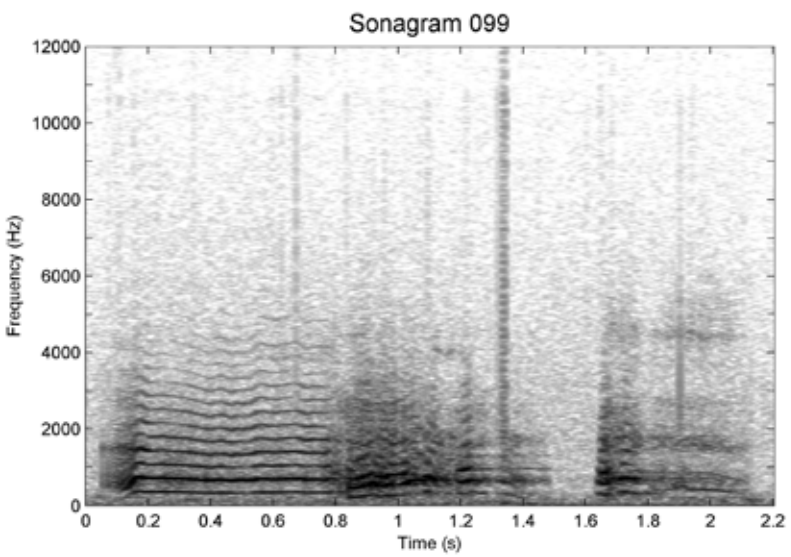

FIGURE 7-101. Congenital, circumscribed anomaly of the tracheal cartilages. Infant aged 17 months. *History:* Frequent episodes of catarrh. "Peculiar" cough. *Laryngoscopy:* Mobile, intact vocal cords; normal mucosa. *Tracheoscopy:* The third and fourth tracheal cartilages, although distinct, are smaller than the rest, are flattened, and protrude into the lumen. *Acoustic finding:* Hollow cough. *Sonagram No. 99:* 0.05 to 0.75 s dull cry with imperfect harmonic structure extending to 4500 Hz only. 0.75 to 1.3 s: Confluent noise up to 5000 Hz, showing dense, overtonelike ribbing corresponding to the protracted, hollow cough. There is a component of increased intensity around 800 Hz.

The third type, that is, circumscribed anomaly, is the least common of the three as shown by our data and also by respective data in the literature. It is a congenital malformation affecting one or two tracheal cartilages which may protrude into the lumen of the trachea.

The symptoms depend on the degree of stenosis. Usually, as also in our cases, they are less severe than in tracheomalacia or rigid stenosis.

KEY POINT

The three most common forms of intramural tracheal stenosis are: (1) tracheomalacia with or without dyskinesis, (2) rigid stenosis, (3) deformity of one or two tracheal cartilages. The third one is the mildest form of the tracheal obstruction. Occurrence and nature of the symptoms vary with the degree of stenosis. Hollow cough, dull cry, and mild stridor may be observed. Tracheoscopy and CT can demonstrate the site and form of the stenosis.

A protracted, hollow coughing sound is the most characteristic auditory finding (Figure 7-101). *The cry may be dull and, depending on the degree of stenosis, stridor may also develop.*

If the pathologic signs described suggest stenosis of the trachea, tracheoscopy is performed which will demonstrate the site and form of the lesion. CT scan can also help in the diagnosis. The therapy depends on the extent of the deformity; it is often only an adjuvant, symptomatic intervention.

Tracheal Dyskinesia

The membranaceous part of the dyskinetic trachea is flabby and shows an increased mobility.

KEY POINT

In infants, the flabbiness and increased mobility of the pars membranacea may be observed even physiologically. It is to be considered pathologic only if it is of extreme degree or it persists in older children. In this latter case a typical hollow cough appears: not the anomaly itself causes the cough, but it makes the cough, which is due to frequent concomitant catarrh, typically hollow. Stridor never occurs. The dyskinesis may be diagnosed by tracheoscopy during expiration (at the end of the examination performed in relaxion).

The caliber of the trachea and bronchi change on respiration (inspiratory dilatation and expiratory constriction) in a purely passive manner due to changes in thoracobronchial pressure. The extent of the caliber changes is determined by two factors, namely, the changes in the extrabronchial intrathoracic pressure and the stability of the tracheal and bronchial walls (Minnigerode, 1972). The wall of the trachea of neonates and infants is less firm than in adults. Especially the flabbiness and increased mobility of the pars membranacea can be frequently observed even physiologically. It is considered pathologic only if it is of extreme degree or persists in older infants. Dyskinesis may be associated with a flattening of the tracheal cartilages (tracheomalacia, functional stenosis, see previous discussion). Minnigerode (1972) distinguished two forms of tracheobronchial dyskinesis: (1) collapse of the trachea, that is, weakness of the cartilages; (2) prolapse of the trachea, that is, atony of the membrane. Tracheal dyskinesia can either be isolated or as-

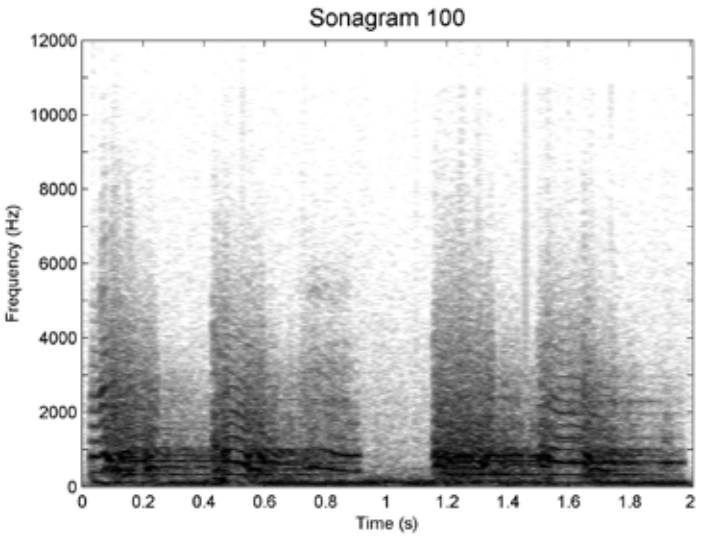

FIGURE 7-102. Dyskinesia of the trachea. Infant aged 12 months. *History:* Caught cold at the age of 6 months, since then frequent episodes of coughing, mainly during the day. *Chest x-ray:* Contraction of the anterior segment on the left superior lobe, with pleural callus. *Laryngoscopy:* No pathologic alteration. *Tracheobronchoscopy:* The pars membranacea of the trachea strongly bulges forward during expiration or coughing, whereas remains unchanged during inspiration. The orifice of the superior lobe of the left lung is narrowed. *Bronchography:* The superior lobe of the left lung and the lingula are supplied by different branches. The cough is due to contraction and pleural involvement resulting from bronchial anomaly. The increased mobility of the pars membranacea makes the cough hollow. *Acoustic finding:* hollow cough. *Sonagram No. 100:* The noise components, which show overtonelike arrangement and are 250 to 300 Hz apart, form an amorphous screen over the entire spectrum. The cough is hollow especially between 1.6 and 1.8 s.

sociated with a more complex syndrome of malformations: esophageal atresia, tracheoesophageal fistula, and laryngotracheal cleft. Although our knowledge of the embryologic development of the tracheoesophageal axis remains limited, the existence of these associations suggests that tracheal dyskinesia is of congenital origin (Corré, Chaudré, Roger, Denoyelle, & Garabédian, 2001).

In dyskinesia of the trachea a typical hollow cough (Figure 7-102) can be observed. In our opinion, increased mobility of the pars membranacea in itself does not cause coughing, but its presence makes the cough, due to the frequent concomitant catarrh (the air passages are narrower and there is a susceptibility to inflammation), typically hollow. Unlike in tracheomalacia, we have not observed stridor or any symptoms suggesting air passage obstruction. Nevertheless, the cry may be hollow.

The diagnosis is established by bronchoscopy made in relaxation, when the spontaneous respiration returns. Sometimes even laryngoscopy can reveal the excessive bulging of the pars membranacea during expiration to an extent which normally occurs only during coughing. The calibre of the trachea is normal in the inspiratory phase; collapse of the posterior membranous portion of the trachea is visible only in expiration. Collapse of the posterior membraneous portion of the trachea during expiration in chronic obstructive pulmonary diseases

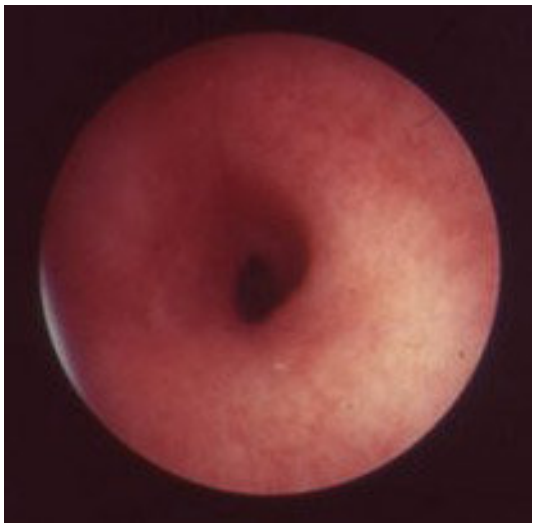

FIGURE 7-103. Endophoto about a rigid stenosis of the trachea in an infant aged 12 months.

in adults and older children is a frequent finding, but *dyskinesis of the trachea is often misdiagnosed as, for example, asthma, thus, to perform an endoscopy, is essential in the evaluation*. Radiologic procedures and pulmonary function tests also may be performed. Management is symptomatic. Tracheal dyskinesia has a good long-term prognosis, as spontaneous resolution is the rule as the child grows up. Very rarely, severe forms and predominating obstruction may be treated by surgery using a connective tissue graft to stabilize the collapsing trachea (Keller & Herzog, 1983).

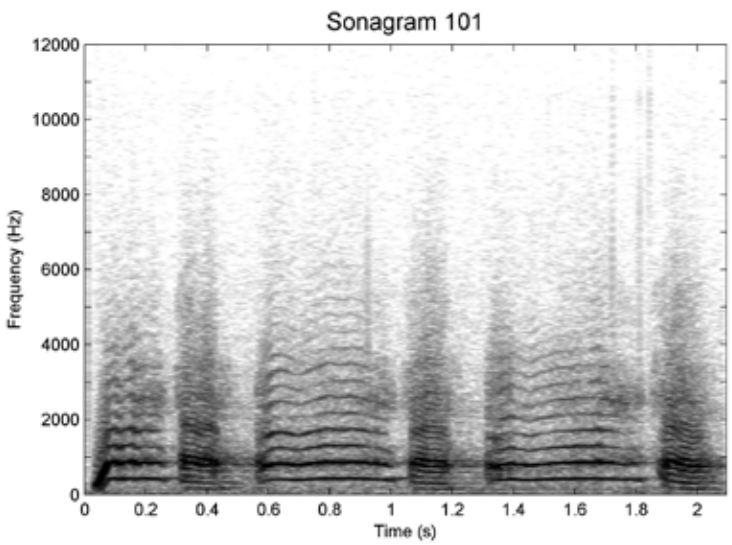

FIGURE 7-104. Congenital rigid stenosis of the trachea (associated with Sedláčková's = VCF syndrome). Infant aged 3 months. *History:* Increasing stridor for one month. Frequent aspiration during feeding. *Laryngotracheoscopy:* Mobile, intact vocal cords. About 1 cm under the vocal cords, in the uppermost segment of the trachea, there is a circular, rigid stenosis covered with normal mucosa. It is only passable by a size 3 bronchoscopy tube. Congestion of exudate underneath the stenosis. *Tracheography:* Approximately 3 cm long stenosis. *Acoustic finding:* Deep, hollow tracheal stridor. *Sonagram No. 101:* 0.05 to 0.3 s, 0.5 to 1 s, and 1.4 to 1.8 s: Normal but dull cry (the acoustic structure is incomplete, extending to 4500 Hz only); 0.25 to 0.45 s, 0.95 to 1.15 s, and 1.7 to 2.0 s: Confluent noise up to 8000 Hz with overtonelike components at about 450-Hz intervals. Around 800 Hz there is an intensive noise component. Hollow, deep tracheal stridor of short duration, which is perceived as a low-pitched sound owing to the low-frequency noise (not visible on the sonagram).

Rigid Tracheal Stenosis (congenital fibrous stricture)

The normally semicircular tracheal cartilages form full rings over shorter or longer portions. The pars membranacea is missing, and the trachea is incapable of dilatation. The congenital fibrous stricture of the trachea may occur as a definite membranous web. Bronchoscopes with external size of 4 or 5 mm, applied normally in neonates and infants, cannot be passed through the rigid stenosis, the lumen of which may be 2 to 3 mm in diameter (Figure 7-103). The severity of the symptoms of respiratory obstruction depends on the size of the lumen.

An excessive inspiratory, or rarely expiratory stridor (which may have tracheal or hollow character) and a hollow, in other cases ringing, metallic, cough (Figure 7-104) *is found. The cry is clear or sometimes dull.*

KEY POINT

Severe respiratory symptoms suggest rigid tracheal stenosis. In this anomaly the tracheal cartilages form a full ring, the pars membranacea is missing. An excessive inspiratory, or sometimes expiratory stridor and a hollow, ringing, metallic cough may be heard. The cry is clear, sometimes dull. Diagnosis can be made by tracheoscopy: the scope with normal measures cannot pass through the rigid stenosis. The length of the obstruction can be determined by tracheography or, rather, with CT/MRI. In severe cases excision of the stenotic part of the trachea and an end-to-end anastomosis is indicated.

Pathologic sounds and the usually severe respiratory symptoms suggest tracheal stenosis with a high degree of probability. Diagnosis can be made by means of tracheoscopy, whereas the length of the stenosis is determined by tracheography or CT scanning (Berrocal, Madrid, & Novo, 2004).

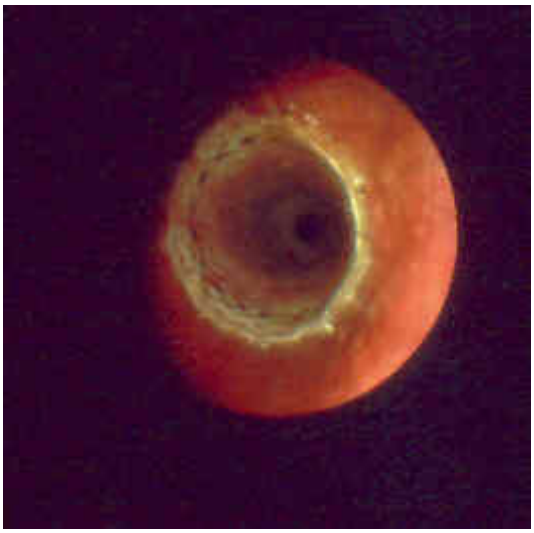

FIGURE 7-105. Stent in the trachea.

In severe cases surgery is indicated: excision of the stenotic part of the trachea and end-to-end anastomosis. Alhough tracheal resection in children for tracheal stenosis is associated with the risk of a higher rate failure than in adults, even severe stenosis and long segment resection may achieve excellent results. Preciado, Cotton, and Rutter (2003) advise that important factors influencing the outcome in these surgeries include anastomosis to an intact cricoid ring, and support of the anastomosis with a tracheal stent. The most important postoperative problem in tracheal surgery is the development of granulation tissue (Schweizer, Berger, Petersen, Kirschner, & Schweizer, 2005). According to some authors, before the surgery a stent may be applied for wid-

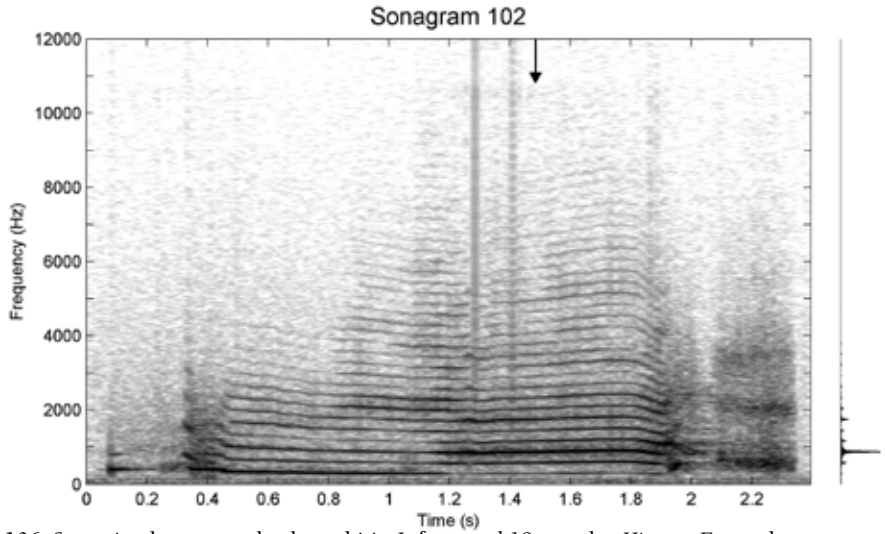

FIGURE 7-106. Stenosing laryngotracheobronchitis. Infant aged 18 months. *History:* Fever, dyspnea, prostration, cough and difficult breathing for 2 days. *Laryngoscopy:* Inflammation of the larynx, swollen subglottic mucosa with a crust and a small amount of dense pus. *Acoustic finding:* Dull, slightly hollow cry; inspiratory stridor; occasionally hollow, deep cough. *Sonagram No. 102:* 0.4 to 1.9 s: Regular, densely ribbed harmonic structure extending first (0.4–0.9 s) to 2500 Hz only, then over the entire width of the spectrum. Noisy thickening of the individual overtones (this is why the cry is not ringing). The ribs about 350 Hz apart (up to 2500 Hz) make the cry hollow, whereas dullness in the first part of the record is due to the low frequency of the overtones; 1.1 to 1.9 s: Regular, loud inspiration.

ening the lumen (Figure 7-105). Others (Clement, Geddes, & Best, 2005; Othersen, Hebra, & Tagge, 2000) recommend endoscopic carbon dioxid laser division of complete tracheal rings which seems to be a safe and effective method of treating congenital tracheal stenosis in exeptional cases, also in infants.

Stenosing Laryngotracheobronchitis (sicca maligna)

This is usually a severe condition with high fever, poor general condition and severe respiratory symptoms caused primarily by virus infection. Sometimes, secondarily, it may be associated with bacterial infection, which, however, in other cases may be the primary etiology (Sie, 2000). As opposed to supraglottic inflammation, it is relatively common in infants. Bacterial tracheitis is now the most common paediatric airway emergency: its incidence has been steadily increasing since 1990, culminating in 2003 (Devlin, Golchin, & Adair, 2007).

The mucosa of the affected areas exhibits purulent, ulcerative changes, with a dense, viscid exudate, obturating crusts, and pseudomembrane; there may be an associated edema of the subglottic area. The diversity of the symptoms is obviously due to the fact that they depend on the agent causing the superinfection (*Pseudomonas*, *pyocyanea*, *Hemophilus influenzae*, *Moraxella catarrhalis*, *Staphylococcus aureus*).

The symptoms and the associated sound changes depend on the location of the fibrinous coat and crusts. In typical cases we hear a veiled or hoarse, occasionally also slightly

hollow or dull cry, subglottic stridor, and a barking or hollow-catarrhal cough (Figure 7-106). *Thus, in addition to dyspnea and stridor, we may find either an abnormal cry or a barking, irritative cough, depending on whether it is the larynx or the trachea and bronchi which is more seriously involved. Massive crusts, tenacious discharge, or fibrin plugs may produce total aphonia* if they (i) are localized between the vocal cords and prevent their closure; or (ii) develop below the level of the vocal cords and obstruct the subglottic lumen. Subglottic pressure required for phonation will be inadequate in this latter case. Severe dyspnea or apnea may accompany aphonia.

KEY POINT

Stenosing laryngotracheobronchitis is a severe condition with high fever, poor general condition, and severe respiratory symptoms. These symptoms depend on the location and degree of the dense, viscid exudate, fibrinous coats, and crusts. In typical cases hoarse cry, subglottic inspiratory stridor, and barking, irritative cough may be present. Laryngobronchoscopy clarifies the diagnosis. Immediate improvement of the symptoms can be achieved by removal of the crusts. Adaequate antibiotic therapy is essential; long-term intubation—because of the possible later consequences as cicatrization—is not recommended.

Based on these general symptoms and acoustic findings laryngobronchoscopy is performed, which clarifies the diagnosis. Lateral airway films may be useful in diagnosis. The tracheal air column may appear diffusely hazy with multiple luminal soft tissue irregularities indicative of pseudomembrane detachment.

FIGURE 7-107. Fibrin moulding of the tracheobronchial tree in a severe fibrinous tracheobronchitis in a child aged two and a half years.

Immediate improvement of the symptoms can be achieved by endoscopic removal of the crusts and aspiration of the exudate which may be considered as an endogen foreign body (Figure 7-107).

Thus, the *bronchoscopy is diagnostic and therapeutic at the same time*. An *adequate antibiotic therapy with broad spectrum of antibiotics is essential*, cold humidity is also suggested. The use of *corticosteroids* in patients with laryngotracheobronchitis was controversial for many years but has, in the last decade, *transformed the management of this disorder* (Stannard & O'Callaghan, 2002). Most children respond to a single, oral dose of dexamethasone. Nebulized epinephrine should be reserved for patients with moderate to severe laryngotracheobronchitis. Simultaneous administration

of corticosteroid and epinephrine reduces the rate of intubation (Leung, Kellner, & Johnson, 2004). According to our experiences, long-term intubation is mostly contraindicated because of the possible later consequences (cicatrization, permanent subglottic stenosis). In these cases we prefer the tracheotomy which is, however, rarely indicated.

Foreign Bodies in Airways

Aspiration of foreign bodies occurs frequently in infants and in young children (Kallay, Hirschberg, & Csermely, 1967). The most variable materials may be asphyxiated (Figures 7-108, 7-109, and 7-110)

The symptom and the sound phenomena observed are most variable; they depend on the size, shape, quality, and position of the foreign bodys as well as their duration (acute or chronic foreign body).

When there is evidence of aspiration in the history, stridor, dyspnea, or cyanosis are unquestionable symptoms of a foreign body.

In addition to the symptoms of suffocation, a foreign body lodged in the larynx in between the vocal cords produces marked

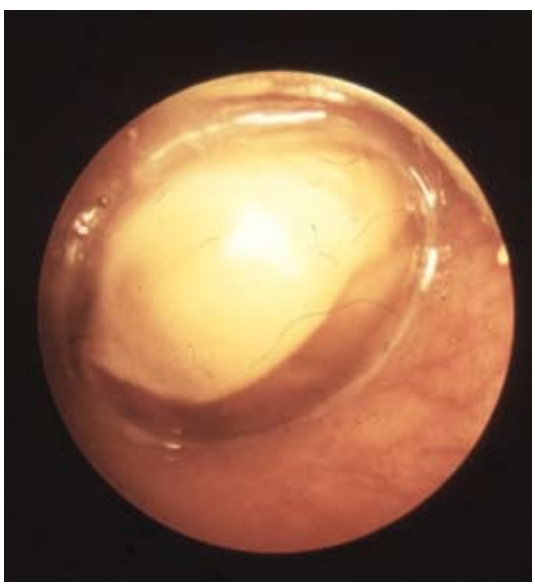

FIGURE 7-108. A bean in the trachea. Infant aged 10 months.

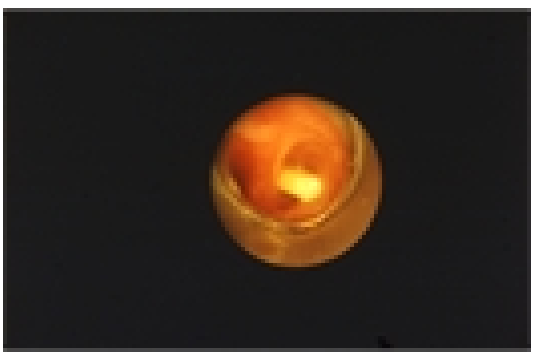

FIGURE 7-109. Nutmeat in the main right bronchus. Child aged 13 months.

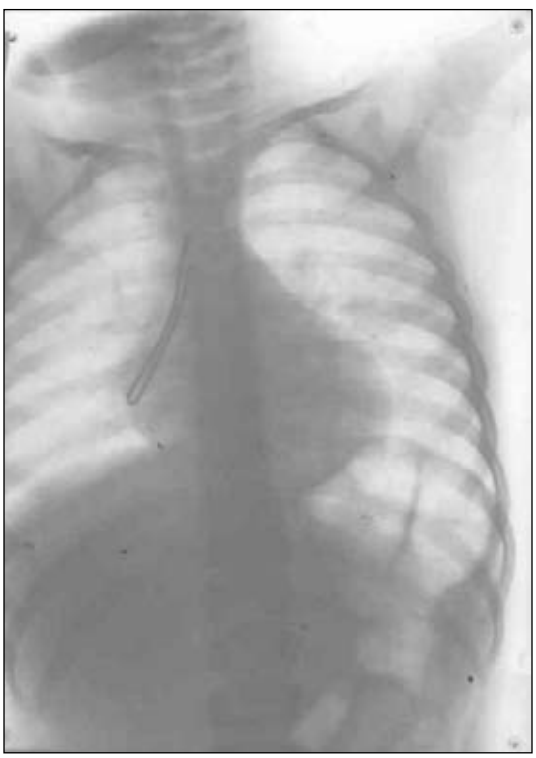

FIGURE 7-110. Hair-slide in the right main bronchus. Infant aged 14 months.

hoarseness or even aphonia. If it is wedged in the subglottis, symptoms resembling those of pseudocroup dominate (dull cry, subglottic stridor, barking cough). A foreign body floating in the trachea may produce an "auditorys thud" which is heard at the end of expiration when the foreign body strikes the undersurface of the vocal cords. A foreign body in the trachea with or without obliterating it may simulate the symptoms of crusty, malignant tracheobronchitis (without fever), that is, an irritative cough and noisy inspiration and expiration. Foreign body in the bronchus may cause suffocation and intensive cough.

According to publications from several parts of the world (Brkic & Umihanic, 2007; Midulla et al., 2005; Ramirez-Figueroa, Gochicoa-Rangel, Ramirez-San, & Vargas, 2005) most of the foreign bodies are organic in nature; vegetables, in the right main bronchus. They may be more dangerous than the inorganic pieces as they can become swollen, increasing the possibility of suffocation. The types of the foreign bodies did not change markedly in the last 50 years (White, Zdanski, & Drake, 2004).

KEY POINT

Aspiration of foreign body occurs frequently in infants and young children The symptoms and the respective sound phenomena observed are most variable: their variance lies in the size, shape, quality, and position of the foreign body as well as in the duration of its presence. Hoarseness, aphonia, stridor, barking cough may be present (detailed in text).When the history, clinical symptoms, or radiologic findings arouse the vaguest suspicion of the presence of a foreign body, immediate laryngobronchoscopy is indicated.

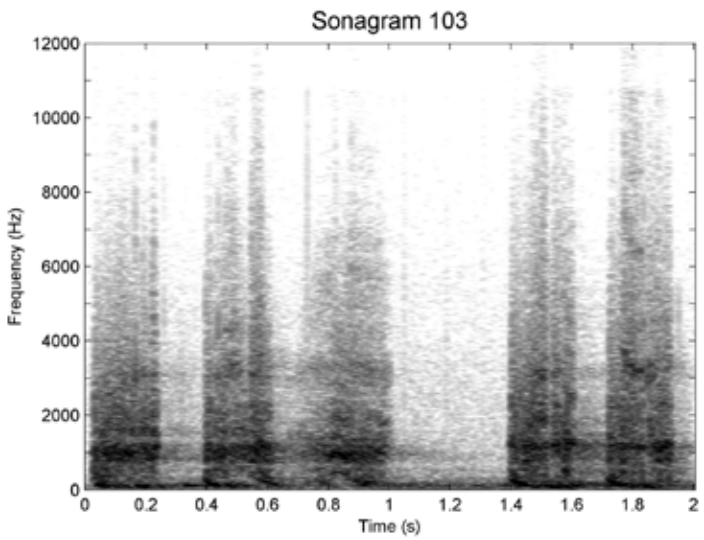

FIGURE 7-111. Foreign body in the trachea and bronchus (a 9-cm long husk, removed). Infant aged 6 months. *History:* Coughing, occasionally paroxysmal, for 10 days; no fever. *Chest x-ray:* Atelectasis of the middle lobe of the right lung. *Bronchoscopy:* Foreign body in the lower one-third of the trachea protruding into the right bronchus; there is a dense, purulent discharge behind it. *Acoustic finding:* Catarrhal cough (with occasional other, highly variable sounds). *Sonagram No. 103:* Confluent noise from 1000 Hz throughout the entire width of the spectrum. The typical record of a dull, catarrhal cough without ringing appears between 1.4 and 1.65 s, and between 1.75 and 1.95 s.

In the majority of cases after the above described initial phase with marked acute symptoms the foreign body lodges in one of the main bronchi, and a silent period follows, the duration of which is variable. Chronic foreign bodies may produce nonspecific symptoms mimicking chronic bronchopulmonary diseases (bronchitis, asthma, pertussis, pneumonia) (Kallay et al., 1968a, 1968b, 1968c). There may be coughing, which is mostly catarrhal (Figure 7-111), but may also be nondescript, or may be accompanied by spastic breathing or a wheeze.

When the history, clinical symptoms, or radiologic findings give rise to the suspicion of the presence of a foreign body, bronchoscopy should be performed. Rigid bronchoscopy is the preferred method for removal of foreign bodies lodged in the airways, but some studies found that flexible bronchoscopy also can achieve a high success rate (Ramirez-Figueroa et al., 2005). Plain chest radiography remains the initial imaging modality for patients with clinically suspected tracheobronchial aspiration (Pinto et al., 2006), except, for very urgent cases, of course. On the other hand, according to the study of Midulla et al. (2005) clinical symptoms and radiologic findings before bronchoscopy have a low diagnostic value. Brkic and Umihanic (2007) are correct that *prevention of aspiration of foreign bodies is better than cure.*

Disorders Affecting the Bronchi and the Lungs

Wheezy Bronchitis, Asthma

Bronchitis is an inflammation of the bronchus usually caused by an infection, whereas wheezing is an acoustic phenomenon which means a high-pitched whistling sound associated with breathing. Wheezing occurs when a child tries to breathe deeply through air passages that are narrowed or filled with mucus as a result of allergy, infection, illness or irritation (Gale Encyclopedia of Medicine, 2006). Wheezing most often comes from the small bronchial tubes, but it may be due to a blockage in larger airways. Common causes are, among others, asthma, bronchitis, bronchiolitis, gastroesophageal reflux disease, viral infection, and pneumonia.

Asthma and wheeze in childhood are complex entities, presenting with different characteristics at different ages, and implicated varied and changing causes (Wright, 2002).

Transient early wheezy bronchitis is generally accepted to define recurrent wheezing in nonatopic babies and tends to disappear by the age of 3 years. The primary risk factor is reduced to pulmonary function since birth and the most common cause is viral infection. Wheezing in infants and toddlers is a common symptom, but not all wheezy bronchitis is, or will become, asthma. Asthma can start at any age and may be provoked by viruses, family history of atopy, and early allergic sensitization (Horak, 2004). Wheezy bronchitis is a separate syndrome of atopic asthma (Christie et al., 1999). Typical bronchial asthma tends to occur in older children. The incidence of severe wheezing bronchitis was 3 in 1,000 children in Sweden (Rylander, Eriksson, Pershagen, Nordvall, Ehrnst, & Ziegler, 1996). As far as the mechanism of wheezing bronchitis is concerned, it is an allergic inflammation with the principal symptoms of spasm in

the small bronchi and dyscrinia (production of excessive amounts of sticky mucus).

KEY POINT

The symptoms of wheezy (spastic) bronchitis are typically: spastic expiratory noise and catarrhal cough. The stridor may be low or high-pitched, sometimes coarse or whistling in character. The clinical picture leaves little doubt as to the diagnosis; one should, however, always consider that behind the symptoms various diseases or alterations could be hidden, for example, foreign body or airway obstruction.

Bronchitis causes nondescript or catarrhal coughing, whereas spasm of the small bronchi elongates the duration of the expiration and brings about a characteristic spastic expiratory noise (Figure 7-112). *This typically expiratory stridor may be high or low pitched, occasionally coarse, whereas in other instances components characteristic of whistling may also appear, presumably due to the viscid exudate. In its abbreviated form it is also called wheezing.*

The typical stridor sound leaves little doubt as to the diagnosis, which is quite obvious from the clinical picture anyway. *One should always consider*, however, *that behind the symptoms*, *seemed to be characteristic* to spastic, wheezing bronchitis, *various other diseases or alterations could be hidden*, as for example, foreign body or airway obstruction. For typical nonatopic wheezy bronchitis a therapeutic trial with bronchodilators is generally sufficient (Horak, 2004).

Pertussis (whooping cough)

Since the introduction of compulsory active immunization pertussis has become a rare

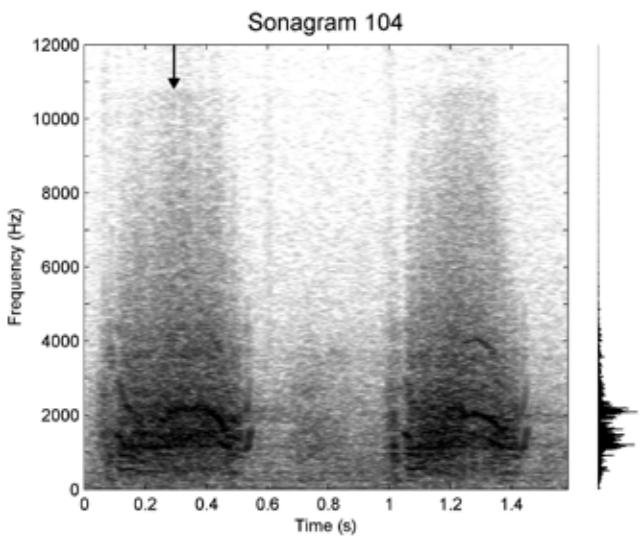

FIGURE 7-112. Wheezy bronchitis. Infant aged 6 weeks. *History:* Dyspnea for 2 days. Prolonged, spastic expiration, marked dyspnea and poor general condition on admission. *Acoustic finding:* Slightly sibilant, spastic expiratory stridor (wheezing). *Sonagram No. 104:* Confluent noise bands up to about 5500 Hz; intensive components at 1500, 2300, 3000, 4000, and 4800 Hz. The high-frequency components (around 3000, 4000, and 4800 Hz) make the stridor sound high pitched.

disease. The number of reported cases is more than 97 to 99% lower than in the prevaccine era (Tanaka et al., 2003), but it is also a fact that the number of cases has been growing again since the 1980s (Marchant, 2005). First, adolescents and adults fall ill, but the atypic pertussis of the adults, however, seriously endangers the infants who are not yet vaccinated (Plotkin, 2005). Infants under the age of 12 months have more serious illness from pertussis and are more likely to have complications (pneumonia, convulsions) than persons in other age groups. The number of cases clinically considered as pertussis yearly is 10 to 20 among infants and children in Hungary with 10 million inhabitants (Budai, 2005). In the United States, among infants aged 5 to 11 months, there was no increase in the reported rate from the 1980s to the present: 89 per 100,000 infants.

In its typical form it has three stages: the catarrhal stage, the paroxysmal stage, and the stage of decline. Coughing is a typical symptom in all three phases. The catarrhal stage begins with a runny nose, sneezing, low-grade fever, and a mild cough which progresses in intensity. At first it is nondescript or catarrhal, whereas by the end of the second week the paroxysmal stage a deep, loud, drawing inspiration is followed by an attack of spastic, staccato coughing (Figure 7-113), which, in turn, is followed by a usually long, shrieking inspiration, stridorphonation, or inspiratory phonation, so-called whoop. The number of attacks varies with the severity of affection. Six or seven, but even 50 attacks may occur daily (Petényi, 1961). *Characteristic is vomiting after the*

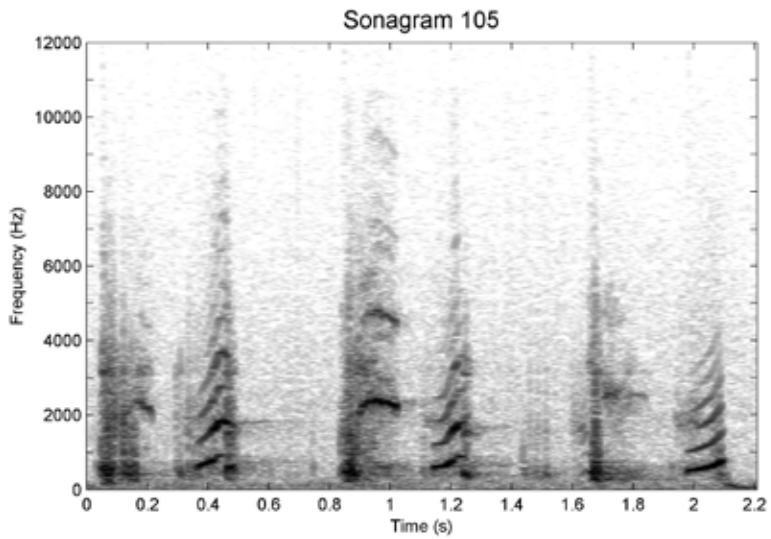

FIGURE 7-113. Pertussis. Infant aged 10 months. *History:* Catarrh with cough for 2 weeks. Paroxysms for a few days. *Acoustic finding:* Staccato cough and stridor-phonation. *Sonagram No. 105:* 0.05 to 0.2 s, 0.8 to 1.0 s, and 1.65 to 1.8 s: Rising and falling overtones between 2000 and 2500 Hz, then at 5000 and occasionally around 7500 Hz within the noise characteristic of the cough; 0.3 to 0.55 s, 1.1 to 1.25 s, and 1.9 to 2.05 s: Rapid and short inspiratory phase following the cough characterized by stridor-phonation having a regular harmonic structure. Due to the scanty overtones of both the staccato cough and the inspiratory stridor-phonation, the sound has a ringing character.

coughs. In the stage of decline (reconvalescence) the paroxysms decrease in intensity and duration, and finally also become less frequent. This is in accordance with the observation of Sewell (1983). According to his description *cough in pertussis is dramatic and very characteristic: a patient with this disease produces a series of short, sharp, staccato coughs rapidly getting closer and closer together in time and increasing in pitch and force*. The main reason for admission to the pediatric intensive care unit of Melbourne Royal Children's Hospital during a period from 1985 to 2004 were apnea with or without cough paroxysm, pneumonia, and seizures (Namachiavayam, Shimizu, & Butt, 2007).

In the paroxysmal stage the diagnosis is evident from the coughing sound. The paroxysmal staccato cough is due to inflammation of the bronchial mucosa leading to the production of glassy, tenaceous mucus, which may become crusty when dry. In the catarrhal stage pertussis may be suspected on grounds of frequent nocturnal coughing and a high leukocyte count with normal erythrocyte sedimentation rate. Isolation and culture of *Bordatella pertussis,* the pathogenic agent, confirms the diagnosis. Unfortunately, the sensitivity of this examination is only 20 to 40% in the first 2 to 3 weeks. After the fourth week agglutinins and complement-fixing antibodies can be found in the blood; their presence, too, is of diagnostic value.

The best way to protect infants from pertussis is to give vaccine in recommended doses. Antibiotics are efficacious solely in the catarrhal stage, but they, especially the macrolids, may be well applied, also later, as chemoprophylaxis, for protection of contact persons (Budai, 2005).

KEY POINT

Since the introduction of active immunization pertussis has become a rare disease. In its typical form it has three stages, that is, the catarrhal stage, the paroxysmal stage, and the stage of decline. Cough is the leading symptom which is first catarrhal, then typical staccato and spastic followed by a usually long, shrieking inspiration (whoop).Characteristic is vomiting after cough. In reconvalescence the paroxysm decreases in intensity and duration. Isolation of Bordatella pertussis confirms the diagnosis.

Pneumonia

In the past 5 to 6 decades there have been radical changes in the approach of pediatry to pneumonia. Whereas earlier it was the classical lobar pneumonia that was most frequently encountered, today bronchopneumonia, that is, focal and interstitial inflammations, predominate. The latter is extremely poor in clinical symptoms, and the evidence obtained by percussion and auscultation is rather indefinite, whereas the radiologic signs may be misleading because of the masking effect of emphysema. In other cases these signs only become demonstrable after the clinical symptoms have subsided. In certain types of pneumonia even the laboratory findings may be normal at the onset. As a result of these diagnostic difficulties which may arise in spite of the advances in laboratory and radiologic techniques, more attention has to be given to the observation of the general condition and appearance of the patient, and to auditory signs if these are also present.

Despite the often minimal physical changes, the infant is in a poor general condition, making the impression of being seriously ill. Breathing is fast and difficult; the expiration

is slightly moaning and requires ancillary muscular activity. *There may be a characteristic, suppressed cough. Due to its painfulness, coughing is composed of phases much shorter in duration than usual* (Figure 7-114).

According to Sewell (1983), in small infants, pneumonia caused by Chlamydia trachomatis may be confused with pertussis: both conditions produce episodes of short staccato coughing and these episodes may be accompanied by apnea and cyanosis. *The cry is clear and regular, but owing to the infant's severe condition it is usually rather weak and moaning.*

The essence of therapy is the application of antibiotics on the basis of the bacteriologic finding. Nowadays cefuroxime is the most often used intravenous antibiotic (Clark, Donna, Spencer, & Hampton, 2007) The finding, management refers to the pediatrician, but the laryngobronchologist may have a role in the improvement of the infant's general condition with the aspiration of the dense secretion (Hirschberg et al., 1970).

KEY POINT

In the past 5 to 6 decades bronchopneumonia, that is, focal and interstitial pneumonia predominate instead of the earlier classic lobar pneumonia. The physical changes and the radiologic signs may be poor, but the infant is in severe general condition. Breathing is fast and aggraviated, the expiration is slightly moaning, and a characteristic, suppressed cough may occur. The management refers to the pediatrician giving the infant antibiotics, but the laryngobronchologist may have a role in the management: with aspiration of the dense secretion (endogen foreign body) he or she can improve the infant's condition.

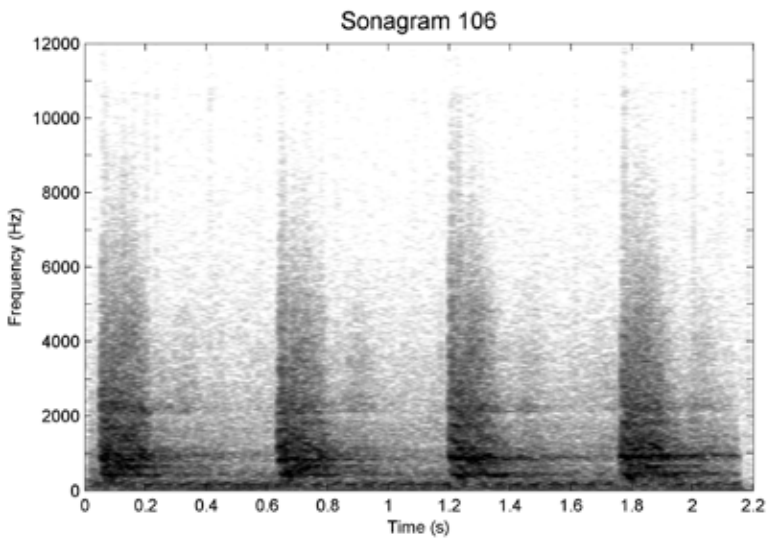

FIGURE 7-114. Pneumonia. Infant aged 3 months. (For the *History* see Figure 6-61) *Acoustic finding:* Suppressed, painful cough. *Sonagram No. 106:* The record resembles that of a nondescript cough, but the noise components are more evenly distributed along the frequency axis and show very little division. There is noise up to about 9000 to 11000 Hz with the dominant intensity level up to 500 Hz. Suppression is indicated by two factors: the relatively short duration of the coughs and the relatively long intervals between them. The short duration "cough pockets" follow each other in a 0.4 s period of time.