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## Submucous Cleft Palate. Diagnostics and Treatment

### SUMMARY

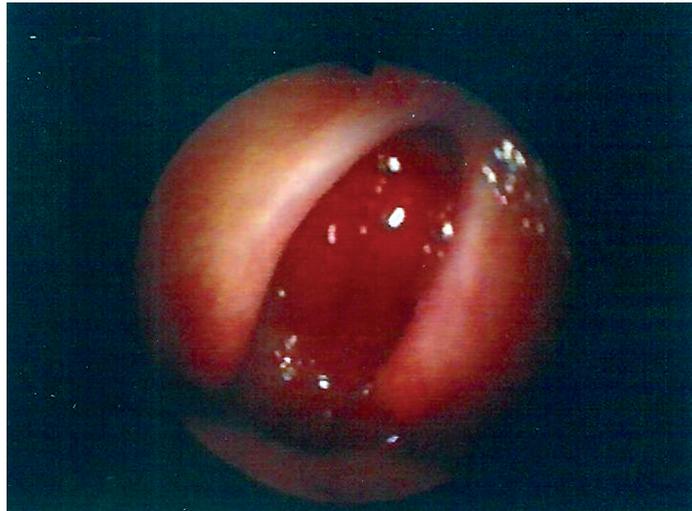
The submucous cleft palate is a specific form of the cleft palate defect, which is highly difficult to diagnose and treat. It may have an overt form, manifesting itself in a characteristic triad of symptoms, and an occult one, which is especially difficult to diagnose. Many of these palatal clefts produce no symptoms. They often become manifested in children after adenoidectomy in the form of unexpected hypernasality of varying intensity after surgery. The study presents basic distinctive features of this type of cleft palate and usefulness of endoscopic diagnostics for making a correct diagnosis. It also discusses indications for surgery and determinants of the need to perform a palate repair before possible logopedic rehabilitation.

**Key words:** submucous cleft palate, occult submucous cleft palate, velopharyngeal insufficiency, hypernasality (open rhinolalia), nasofibroscopy.

In the pharyngeal phase of swallowing, in yawning, middle ear ventilation through Eustachian tubes, and during the realization of oral sounds it is necessary to entirely separate the oral cavity and middle pharynx from the nasopharyngeal cavity. This separation is determined by the proper velopharyngeal closure. The velopharyngeal sphincter consists of musculature of the soft palate and of the posterior and lateral walls of the throat. The muscle layer of the soft palate is made up of five muscular pairs: the velar elevator muscle, the velar tensor muscle, the velopharyngeal muscle, the palatoglossal muscle and the uvular muscle. The muscle layer of the posterior and lateral walls of the throat is the cephalopharyngeus (superior constrictor muscle of pharynx). The most frequent cause of velopharyngeal insufficiency and the concomitant resonance disorders in the form of hypernasality is the cleft palate.

THE CLEFT PALATE OCCURS IN ISOLATION OR WITH THE CLEFT LIP AND CLEFT ALVEOLAR PROCESS THIS IS THE MOST FREQUENT DEVELOPMENTAL DEFECT IN THE FACIAL SKELETON. IT DEVELOPS BETWEEN THE 4<sup>TH</sup> AND 12<sup>TH</sup> WEEK OF FETAL LIFE. IN POLAND, DEPENDING ON THE DEMOGRAPHIC SITUATION, 600-800 CHILDREN WITH THIS DEFECT ARE BORN EVERY YEAR, AT THIS FREQUENCY RATE: FROM 1.6 DO 2.2/1000 OF LIVE BIRTHS (FIG.1).

1a



1b





Fig. 1. Examples of overt cleft palates: *total isolated cleft soft and hard palate* Partial cleft of the soft palate, *ISOLATED BIFID UVULA*

A special form of the cleft palate defect is the submucous cleft. Unlike overt cleft palates it does not manifest itself after birth in an easily seen “hole”, which even a midwife can notice. In addition, the submucous cleft does not cause feeding difficulties typical of overt palatal clefts. Therefore, on account of its more occult form it causes considerable diagnostic and thereby therapeutic difficulties. Moreover, many of these palatal clefts do not exhibit speech disorders in the late period; consequently they often remain undiagnosed during the rest of life and do not need treatment.

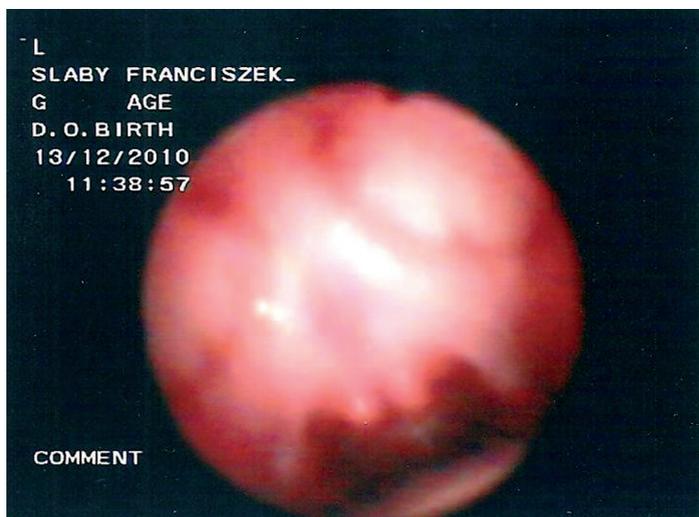
Unlike classic forms of the cleft palate characterized by the midline cleft both in its muscle layer and the mucous membrane, in submucous clefts of the palate, the continuity of the palatal mucous membrane is maintained, both from the oral cavity and from nasal cavities. However, the soft palate muscles covered with it, instead of fusing horizontally in the midline as the palatine suture, are cleft to varying degrees. The submucous cleft palate may also cover the hard palate to different degrees. Muscle fibers therefore run diagonally and depending on the extent of the cleft they hook on the posterior palate margin or along the cleft line in the hard palate, which, in the total submucous cleft palate, may reach as far as the incisor foramen.

The extent of submucous cleft palates thus varies – from the partial diastasis of palatal musculature to the submucous cleft of the whole hard (bone) palate and soft (muscle) palate. They may have an overt or occult form. It should be clearly emphasized that this range of the submucous cleft palate does not go hand in

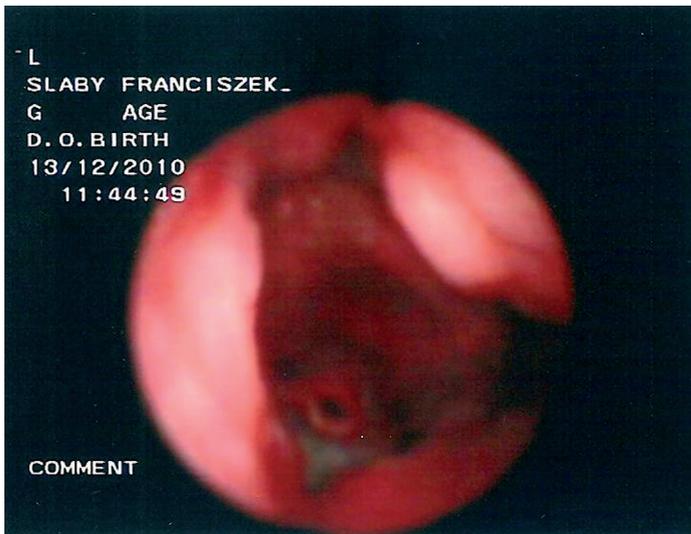
hand with the degree of intensity of speech disorders. The overt submucous cleft palate, easy to diagnose on the intraoral examination, may have no symptoms at all whereas its occult form can be accompanied by gross hypernasality impeding speech intelligibility to a large extent. For the cleft palate symptoms to occur, more significant is the extent of the defect in the purely anatomical sense. Prior to the introduction of endoscopic diagnosis to assess the structure and function of the velopharyngeal sphincter, most occult palatal clefts were diagnosed as the congenital short palate or palatal dysplasias. It was only Croft who demonstrated that the characteristic incisure in the contour of the posterior soft palate margin in the endoscopic image or a considerable flattening of the contour in the bulge area of the uvular muscles, is proof of the submucous cleft muscles, even if there are no other characteristics of the submucous cleft palate on oral examination in the form of Calnan's triad (Ch. Croft et al., 1978).

The overt submucous cleft palate is characterized by the triad of symptoms, first described by Calnan in 1954. It consists of 1) bifurcation of the uvula, 2) translucent zone along the midline of the palate due to the separation of velar musculature and thinning of the submucosa, 3) the presence of a palpable smoothness or a notch in the hard palate in the area of the posterior nasal spine As has been said before, in extreme cases the whole hard palate may be submucously cleft, then the thinning of the whole soft and hard palate in the midline may be observed. (Fig. 2).

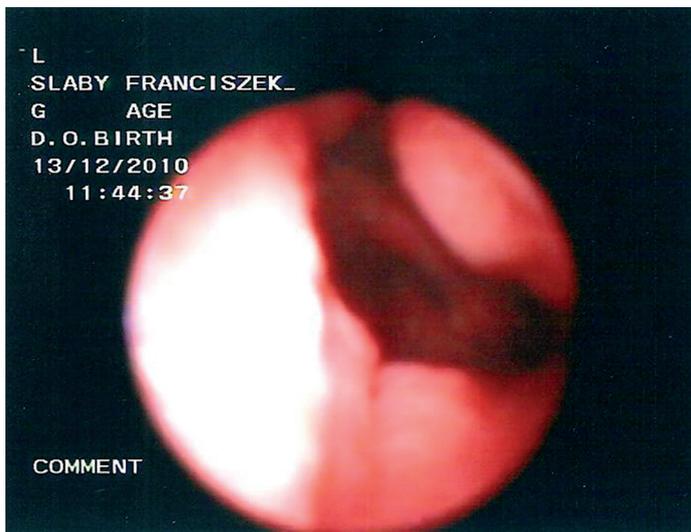
A special form of submucous clefts is the submucous cleft soft palate



2a



2b1



2b2

Fig. 2. The picture of overt submucous cleft palate with a characteristic triad of symptoms: *picture on oral examination* Endoscopic image: a characteristic notch in the posterior border of soft palate is noticeable: b<sup>1</sup>) respiration, b<sup>2</sup>) absence of velopharyngeal closure;

(E. Kaplan, 1975), total or partial. Because of its occult character it is extremely difficult to diagnose, especially on oral examination. The essence of submucous cleft palate is the lack of normal midline fusion of velar muscles with the absence of the above-described symptoms triad typical of the overt submucous cleft. The

total submucous cleft palate involves all the five pairs of soft palate muscles: the velar elevator muscles, velar tensor muscles, velopharyngeal muscles, palatoglossal muscle and the uvular muscle. In the submucous cleft palate, the muscle cleft affects only some part of muscle fibers. For speech functions, what matters only is the possible cleft of soft palate elevator muscles, which then hook abnormally on the posterior border of the hard palate instead of on the palatine suture in the palatal midline. The soft palate elevators are mainly responsible for the palatal movement backwards and up during swallowing, phonation, yawning, and conditions the separation of the oral from nasal cavity during these actions. In contrast to its name, however, the tensor muscle of the soft palate not so much tenses the soft palate as opens the Eustachian tube, when contracting normally. It is therefore necessary for the tube's proper functioning, which conditions the undisturbed ventilation of the middle ear, and thereby proper hearing.

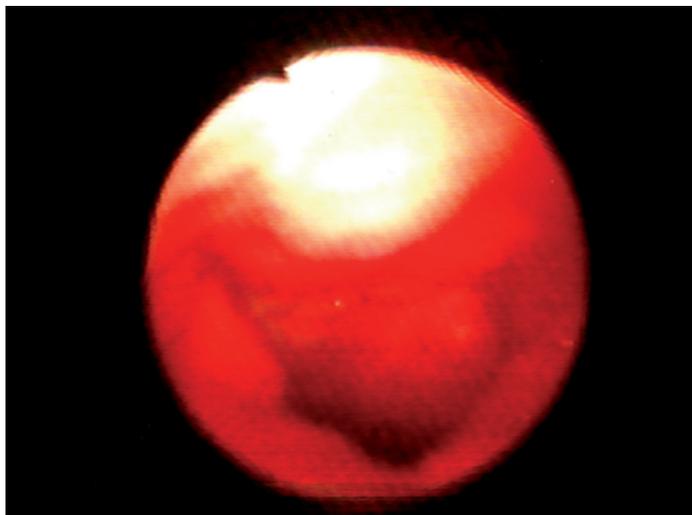
In studies by Stewart, Otto and Lagace (1972) the average frequency of incidence of the submucous cleft palate in the school children population was 1: 1200 of the studied children. However, this frequency is difficult to estimate because many of these clefts, especially those not manifested in resonance disorders, are never detected, as has been said above. It should be added that the bifid uvula occurs in 1% of the "normal" population without any accompanying symptoms of velopharyngeal dysfunction. (R. Weatherley-White et al. 1972) and in itself it does not establish diagnosis of the submucous cleft palate although this undoubtedly requires detailed diagnostic management in order to exclude it, especially before planned adenoidectomy (R. Shprintzen et al., 1985).

With frequently negligible clinical symptoms, a significant additional problem in diagnosing submucous clefts is the lack of sufficient ability to distinguish by ear the character of vocal resonance disorders, i.e. hyponasality (closed rhinolalia), most often caused in children by hypertrophied adenoids or by chronic allergic rhinitis, may not be distinguished from hypernasality accompanying velopharyngeal insufficiency. Sadly, this is the case not only with general practitioners or pediatricians but also with laryngologists, another difficulty being that velopharyngeal insufficiency is often masked by the hypertrophied adenoid. Hence, as the author's practice shows, many a time the first specialist to suspect the submucous cleft palate is the logopedist, who correctly associates the heard hypernasality with the cleft palate defect or velopharyngeal insufficiency of different origin. Unfortunately, in such cases it is often a delayed diagnosis from the standpoint of the capability of repair mechanisms in the speech apparatus, first of all because as a rule children are, regrettably, referred to the logopedist too late.

An additional difficulty is the fact that in children even with slightly hypertrophied adenoids, the velopharyngeal closure is essentially a veloadenoidal one. (S. Gereau and R. Shprintzen, 1988; Y. Finkenstein et al., 1996; Hortis-Dzierzbicka, 2004), while the period of early childhood is the peak period for hypertrophy of

the adenoid, whether within the physiological limits or its considerable hyperplasia. The highly hypertrophied adenoid impedes nasal respiration and ventilation of the middle ear because it blocks the pharyngeal openings of auditory tubes. It is often removed by adenoidectomy for that reason. But in the presence of the submucous cleft, a hypertrophied adenoid may be a kind of “prosthesis” for the velopharyngeal closure, and then its removal may cause the appearance of hypernasality, often in a highly intense form, difficult to accept by the child and his environment. It is for that reason that the child’s palate qualified for adenoidectomy should be always very carefully assessed to exclude the presence of the submucous cleft palate, and in respect of a previous palate operation. Witzel et al. (1986) report that as many 27% of children referred to them for hypernasality, which appeared after removal of the adenoid, were diagnosed with the submucous cleft palate, which evidences the frequency of the phenomenon and thereby the significance of the problem. Care is all the more necessary that as many as 90% of submucous clefts show no speech disorders on condition that no laryngological surgery is performed within the adenoidal system of the throat, especially on the pharyngeal tonsil.

*DIFFERENTIAL DIAGNOSTICS OF OCCULT SUBMUCOUS CLEFT PALATE COVERS CONGENITAL SHORT PALATE, HARD PALATE FOUND IN E.G. THE TREACHER-COLLINS SYNDROME, AND VELOPHARYNGEAL DISPROPORTION. VELOPHARYNGEAL DISPROPORTION OCCURS IN PATIENTS WITH SO-CALLED DEEP THROAT WITHOUT ANY PATHOLOGIES*



Ryc. 3. Ukryty rozszczep podśluzówkowy podniebienia – obraz endoskopowy: charakterystyczne wcięcie w miejsce wypukłości mięśni języczka w obrysie podniebienia miękkiego.

*WITHIN THE PALATE.* THE CONTENT OF NASOFIBROSCOPIC EXAMINATION IN FINAL DIAGNOSIS IN THIS CASE WAS FIRST DESCRIBED BY CROFT (*CROFT ET AL., 1978*). IN NASOFIBROSCOPY INSTEAD OF THE UVULAR MUSCLE BULGE A NOTCH IS OBSERVED IN THE SOFT PALATE CONTOUR IN THE CASE OF THE OCCULT SUBMUCOUS CLEFT. ZDJĘCIE

There are many controversies over referral for surgery in submucous cleft palates and over the procedure itself, given, as has been said above, that many of them show no speech disorders; consequently, they even often go undiagnosed. By contrast, in cases manifesting itself in hypernasality, i.e. exhibiting velopharyngeal insufficiency, indications for surgery are self-evident. Frequently, however, instead of a palate repair operation, corrective operations of velopharyngeal closure are performed as pharyngoplasties (F. Abyholm, 1976; S. Park et al., 2000; M. Abdel-Aziz, 2007). This is, the author and others believe (J. Pensler and B. Bauer, 1988; B. Sommerlad et al., 2004), an entirely wrong practice, because this kind of procedure does not repair the palatal defect. Therefore it does not treat the cause of the diagnosed dysfunction but its effect. Therefore, it is the cleft that should be operated on in the first place, and only then, when this procedure has not produced the expected result despite speech rehabilitation, a child can be referred for pharyngoplasty. This applies especially to those cases of velopharyngeal insufficiency where the submucous cleft has been diagnosed early.

It should be remembered that long-lasting logopedic rehabilitation of speech disorders in unoperated submucous cleft palates is a case of malpractice since the existing structural disorder determines its inefficacy. Another case of malpractice is also to perform only a corrective operation of the velopharyngeal closure in this type of cleft, without first joining the muscles of the palate, which, regrettably, still occurs (M. Abdel-Aziz, 2007). Pharyngoplasty alone, leaving the muscular opening characterizing the cleft, should be limited exclusively to selected cases of velopharyngeal insufficiency without a cleft. However, because of the frequently late surgical closing of the submucous cleft, caused by its late detection and the accompanying hypoplasia of unused muscles, in many cases, simultaneous with a palate operation, pharyngoplasty is performed using the velopharyngeal flap. For, as demonstrated by Hussein et al. (2004) and confirmed by our clinical observations, the results of surgical treatment of submucous clefts are as a rule worse than in overt clefts. In submucous clefts, even exhibiting hypernasality from the beginning, the average age for palatal surgery is 5-6 years. The critical period for the correct speech development has thus been considerably exceeded in these cases (M. Hortis-Dzierzbicka and E. Stecko, 2005)

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