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REVIEW ARTICLE

Acute isolated velopharyngeal insufficiency in children: case report and systematic review of the literature

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Abstract Acute isolated velopharyngeal insufficiency (VPI) is a clinical entity mainly reported in children. We undertook a systematic review in order to better characterize its features. Following a Medline search (1960–2012), the authors reviewed and analyzed the cases of acute VPI in children; 36 cases were found. The most common presenting features were hypernasal speech (97 %), nasal reflux (73 %), and dysphagia (49 %). 73 % of the children were males and 27 % females, of 8.9 ± 2.5 years. In all the cases the VPI was unilateral. One quarter of the children had a recent episode of febrile illness and 11 % of the children had an identified infection at the time of presentation (HAV, parvovirus B19, measles, and Coxsackie virus). No associated cause was found in the other cases. All cases resolved completely (67 %) or partially (33 %) without any treatment (89 %) or with prednisolone (11 %). Acute VPI represents a separate entity within the spectrum of VPI and it is a benign self-limiting disorder. The cause remains undetermined but an infectious disorder may play a role at least in some cases. Follow-up is mandatory in order to eliminate progressive conditions such as brainstem neoplasms or inflammatory diseases.

Keywords Velopharyngeal insufficiency · Acute onset · Children · Cranial neuropathy

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Introduction

The velopharyngeal complex is anatomically limited by the soft palate and the lateral and posterior oropharyngeal walls. It represents a functional port between the oropharynx and the nasopharynx, therefore playing an essential role in deglutition, nasal breathing, and speech. Such functions are achieved through the coordination of the following groups of muscles: tensor veli palatini (pair), levator veli palatini (pair), musculus uvulae (single), musculus palatoglossus (pair), musculus palatopharyngeus (pair), and the superior pharyngeal constrictor (single) [1].

Velopharyngeal insufficiency (VPI) results in nasal regurgitation and nasal speech (rhinolalia), and could participate in otologic disease in certain subgroups of patients [2].

Velopharyngeal insufficiency (VPI) is most frequently seen as a congenital condition, often associated with other anomalies. It is common in patients with cranio-facial malformations like palatine clefts, hemifacial microsomia or hemipalatal hypoplasia, trisomy 21, hereditary myopathy, or in CATCH 22 spectrum disorders.

Acquired VPI can be seen after adenoidectomy or tonsillectomy, especially in patients with submucous clefts. Progressive new-onset VPI is most often seen in association with pontocerebellar or brainstem expansive lesions (neoplastic or infectious), ischemia, or demyelinating diseases [3]. It can also be an associated feature in a wide range of neuromuscular diseases or other childhood syndromes such as dystrophinopathies (especially Duchenne's disease), or Moebius syndrome (which associates paresis of multiple cranial nerves) [4].

In opposition to these forms of VPI, sudden-onset VPI is less frequently reported, almost exclusively in children [5–8].

The goal of this review on acute VPI in children is to outline the existence of this particular form of VPI, and to

characterize its clinical features as well as to attempt gaining some insight on aetiology. We present an illustrative case.

Methods

Systematic review

Medline was used as main search engine with combinations of the terms "soft palate", "palsy OR paralysis", "velopharyngeal (insufficiency OR inadequacy OR incompetence OR dysfunction)", and "children OR infant". We screened the abstracts of all the articles and the full text of the relevant ones. References of the latter were also screened in order to obtain further articles.

We included all cases published until January 2012, describing forms of VPI which fulfilled the following criteria: acquired, acute onset, in pediatric patients (age <16 years), and without associated neurological findings. Besides the articles produced by keyword matching, we excluded cases of VPI that were congenital and/or associated with malformations. Language restriction was made for articles written in English, French, German, Italian, and Spanish.

Data extraction

Data of each included article were extracted independently by two of the authors (VW and LN). Discordant findings were resolved through discussion. We retrieved the following variables when available: age, gender, clinical presentation, complementary investigations, treatment, follow-up, and outcome.

Case report

A 9-year-old girl without a relevant medical past presented with sudden voice modification that appeared when she woke up the same day. The child herself noticed that she could not blow through a straw and reported her voice was almost normal when she pinched her nostrils. She did not complain of nasal reflux. The rest of the systematic anamnesis was contributory for an episode of rhinitis that resolved spontaneously a week before symptoms' onset.

Physical examination revealed right-sided unilateral velar paralysis with open rhinolalia (hypernasal speech) and left uvular deviation on phonation (Fig. 1a, b). No skin rash or other neurological focal signs were present. The gag reflex was preserved, and pharyngo-laryngeal and lingual mobility were normal. The rest of examination was non-contributory. Taste-strip testing was normal.

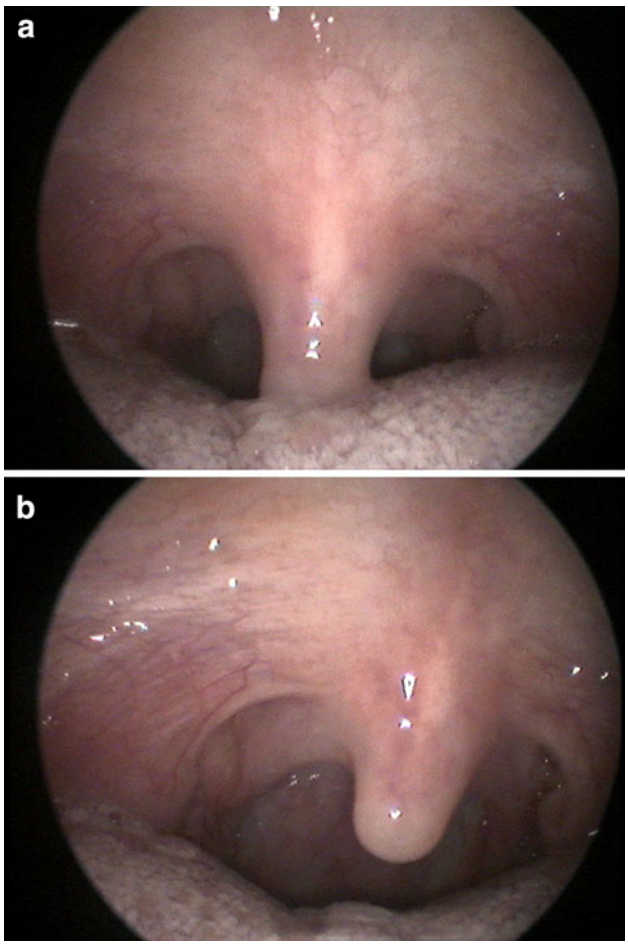

Fig. 1 **a** Soft palate at presentation. No obvious asymmetry can be seen at rest. **b** Velar shift towards the left side during contraction (on phonation), due to right hemivelar palsy

Nasometry, a non-invasive technique used to measure the nasal air leak for non-nasal sounds, revealed a mean nasal air leak of 60 ± 7 % for non-nasal speech (the norm established at our Unit of Speech and Swallowing Disorders for French language speakers is ≤30 %).

Magnetic resonance imaging of the brain was normal, especially the intracranial portion of cranial nerves IX and X.

Serologies were obtained for cytomegalovirus, herpes virus 1 + 2, varicella-zoster virus, hepatitis A virus, and *Borrelia burgdorferi*. They showed a slightly increased value of IgG against *B. burgdorferi* with IgM values within a normal range. Lumbar puncture was recommended but the parents refused this test.

After infectious diseases consultation the patient was treated by doxycycline for 2 weeks, as well as speech therapy. Full recovery was seen at 3 weeks follow-up (Fig. 2), and the nasometry was normalized by that time.

The 1-year follow-up period was uneventful.

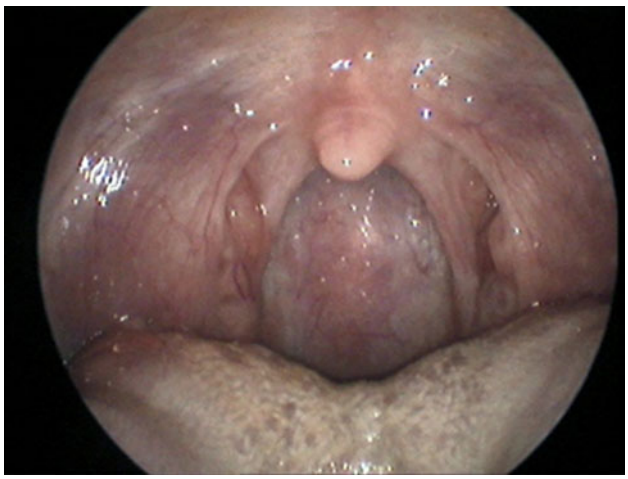

Fig. 2 Symmetric velar contraction 3 weeks after symptom's onset

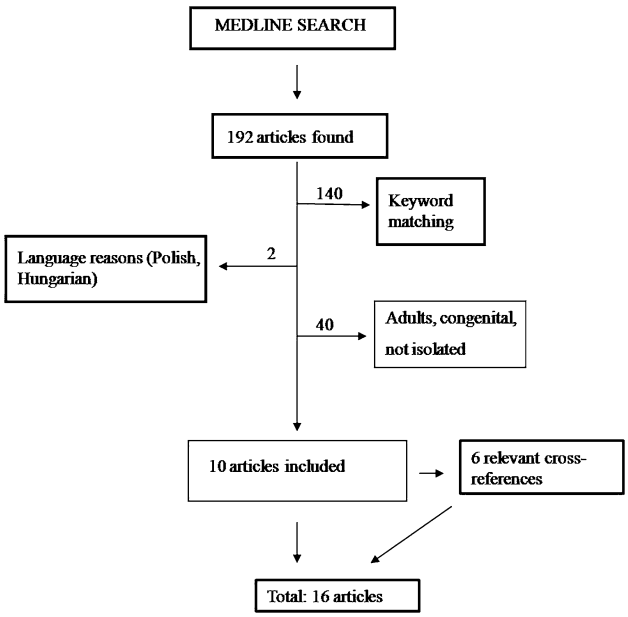

Fig. 3 Flow-diagram with screened, included and excluded articles

Results

Altogether, 192 articles were screened. Of these 176 did not meet the inclusion criteria. Sixteen articles [5–20] describing 36 cases were finally included (Fig. 3). To these we added our case for analysis purposes. Table 1 summarizes patients' features.

All the reports concerned children with a mean age of 8.9 ± 2.5 years old (range 2–18). Acute VPI displays a male predominance as 73 % of the children were male. None of the children had previously known co-morbidities. In all the cases only unilateral involvement was seen, as the only physical finding.

Clinical presentation consisted of new-onset hypernasal speech (97 %), nasal reflux (73 %), and dysphagia (49 %).

Table 1 Patients' features ($N = 37$)

Feature	N (%)
Gender	
Male	73
Female	27
Clinical presentation	
Hypernasal speech (rhinolalia)	97
Nasal reflux	73
Dysphagia	49
Recent fever	25
Transitory dyspnoea	5
Odynophagia	3
Headache	3
Imaging studies	
Plain X-ray	40
MRI	35
CT-scanner	5
Positive findings	0
Microbiological studies	
Serology	60
CSF	22
Stool	3
Positive findings	10
Outcome	
Full recovery	65
Partial recovery	35

Rarer manifestations included odynophagia (3 %), headache (3 %), and short dyspnoeic events (5 %).

A recent history of febrile illness was reported in 25 % of the cases.

Serologies were obtained in 62 % of the cases and 11 % of these allowed documentation of an active infectious context: hepatitis A virus, measles, parvovirus B19 and Coxsackie A9. Table 2 summarizes the microbiological studies performed and their findings.

Imaging of the central nervous system was obtained in 80 % of the patients: magnetic resonance imaging (35 %), computed tomography scanner (5 %), and plain radiography (40 %). Radiological studies failed systematically to reveal any anomalies.

Concerning treatment, 89 % of the patients did not receive any treatment at all, and 11 % were given prednisolone (oral or intravenous).

The outcome was as follows: 65 % of the patients made a full recovery, and 35 % recovered partially (structural and/or functional deficit remaining) within a period between 1 week and 2.5 years after the diagnosis. The mean follow-up time was of 61 months (2 months–20 years); a third of the patients had a follow-up superior to 1 month after

Table 2 Microbiological studies

Pathogen	Tested (*n*/37)	Sample	Positive results
Influenza A + B	10	Serology	0
Parainfluenza	10	Serology	0
RSV	9	Serology	0
Measles	1	Serology	**1/1**
Adenovirus	9	Serology	0
HSV 1 + 2	19	Serology and CSF	0
CMV	16	Serology and CSF	0
VZV	11	Serology and CSF	0
EBV	8	Serology and CSF	0
HHV 6	3	Serology and CSF	0
Enterovirus 1 + 3	1	CSF	0
Echovirus	10	Serology	0
Coxsackie A9	10	Serology and stool	**1/10** (stool)
Poliovirus	8	Serology	0
Parvovirus B19	2	Serology	**1/2**
HAV	2	Serology	**1/2**
HCV	1	Serology	0
Mycoplasma pneumoniae	8	Serology and CSF	0
Chlamydia pneumoniae	4	Serology	0
Queensland spotted fever	1	Serology	0
Coxiella burnetii	1	Serology	0
Legionella spp.	1	Serology	0
Mycobaterium tuberculosis	1	Tuberculin testing	0
Borrelia burgdorferi	8	Serology and CSF	**1/8** (IgG)*

Bold values indicate positive microbiologic findings

CSF cerebrospinal fluid, *RSV* respiratory syncytial virus, *HSV* herpes simplex virus, *EBV* Epstein-Barr virus, *HHV 6* human herpesvirus 6, *HAV* hepatitis A virus, *HCV* hepatitis C virus

* This result corresponds to the case we report

recovery. The length of follow-up was not reported in 40 % of the cases. During this follow-up period no additional co-morbidities were diagnosed in any of the children.

Discussion

This systematic literature review on acute VPI in children reveals that: (a) it presents with stereotypical signs consisting of hypernasal speech, nasal reflux, and dysphagia; (b) unilateral VPI is the characteristic finding in physical examination; (c) concomitant or previous infectious episodes are found in a substantial proportion of the children; (d) imaging studies are non-contributory in the diagnosis; (e) outcome is most often favourable with or without treatment, but one-third of the patients have persistent insufficiency.

The motor supply to the velopharyngeal muscles is mainly provided by cranial nerves IX and X, but the lesser palatine nerve, a branch of cranial nerve VII, also plays a role in the motor innervation of the soft palate. An important feature in the motor velopharyngeal innervation is the interindividual variations in the nervous supply [21, 22].

An important aspect is the characterisation of the lesional level in this particular form of VPI. Based on the findings of this review the answer to this question is not clear. Direct visual transoral examination of the oropharynx combined with fiberoptic nasopharyngoscopy allows precise characterisation of the muscles involved in VPI, but not of the involved cranial nerves. One could speculate that acute VPI could be the manifestation of a transient cranial neuropathy affecting either cranial nerve IX or X.

As can be seen in the results section and as discussed below, imaging studies were not contributory in terms of diagnosis. Nevertheless none of the patients underwent functional imaging and notably functional MRI, but it is possible that such imaging techniques could allow gaining some insight in the cases of "idiopathic" acute VPI.

The cause of acute VPI remains to be elucidated, but one of the most interesting points outlined by the present review is the fact that 25 % of the children presented a concomitant or recent febrile episode. As can be seen in Table 2 microbiological studies were performed in a substantial number of patients. Successful identification was only achieved in four cases. The pathogens were invariably viruses. Three of these are neurotropic viruses (Coxsackie, parvovirus B19, and paramyxovirus), and therefore neurological involvement could be explained by direct viral aggression. In the case of hepatitis A virus, a virus with no known neurotropism, an alternative mechanism to explain neurological involvement is needed. The main hypothesis would be an immunologically mediated response. Indeed, infective or post-infective neurological disease has been reported in the context of hepatitis A [8].

Concerning enterovirus, measles and parvovirus B19, the formation of epitopes with the potential to mediate an autoimmune response after the reactivation of the latent virus (in the neural cells), has been proved experimentally. This could be a second potential mechanism explaining neurological involvement [6, 23].

In line with this potential infectious aetiology the case we report raises the question of a cause-effect relationship between acute VPI and *Borrelia burgdorferi*, one of the causative agents of a tick-borne zoonosis known as Lyme disease or borreliosis.

The neurological manifestations and particularly cranial neuropathy associated with Lyme disease are most commonly seen in the early disseminated stage, while encephalitis, encephalomyelitis and radiculoneuritis are the

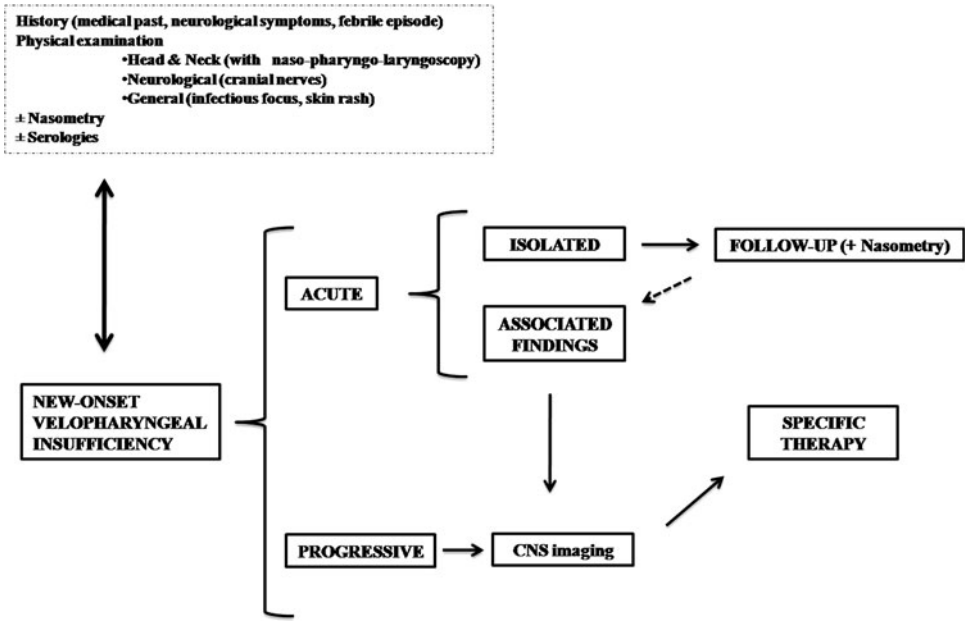

Fig. 4 Suggested approach and management algorithm for new-onset VPI. Follow-up should be performed weekly until recovery

predominant manifestations of late-stage borreliosis [24]. In Switzerland, positive serologies were found in 3–6 % of the general population in the 90s. In geographic areas at high risk, as many as 35 % of the population had positive serologies but only 3.5 % developed consistent clinical symptoms after 10 years [25]. Although the diagnosis of neuroborreliosis could have been eventually confirmed by polymerase chain reaction identification of *B. burgdorferi* in the cerebrospinal fluid, our patient's history, presentation and ulterior evolution were rather inconsistent with this diagnosis. The fact that an improvement was seen under doxycycline does not allow drawing any conclusions on the effect of this treatment, since as this review shows two-thirds of the patients made a spontaneous recovery.

This review outlines acute VPI as an individual entity within the spectrum of VPI. The initial approach of the child with VPI should allow distinguishing between congenital and new-onset VPI. In this last group a thorough history should be obtained including active co-morbidities, trauma, previous surgery, recent or intercurrent febrile episode, and other associated neurological symptoms. Complete head and neck and neurological examination must be performed to assess the unilateral versus bilateral character of the functional velopharyngeal deficit and to identify any associated anomalies.

The acute versus progressive evolution is a feature of utmost importance, as progressive VPI is likely to mirror an underlying condition such an expansive intracranial lesion or a demyelinating disease [26].

The authors of the two first reported cases in 1976 referred to this entity as "isolated temporary pharyngeal paralysis in childhood" [19, 20]. The isolated character is also extremely important since this element could help the clinician to guide the diagnostic workup and the follow-up modalities.

Therefore we propose the term "acute isolated velopharyngeal insufficiency" for the forms of acquired VPI that have the following features: sudden onset without associated neurological findings and unilateral.

If these criteria are fulfilled, the data retrieved from the literature shows this subtype of VPI is a benign and self-limiting condition, and that it does not represent the manifestation of a serious underlying condition. This statement is further supported by the fact that imaging studies of the central nervous system, notably CT-scanner and MRI, were systematically normal. Based on these premises a wait-and-watch attitude could be recommended if neurological examination and notably cranial nerve examination is reassuring. The value of serologies in this context is uncertain, but microbiologic documentation could be attempted when a consistent history of recent or concomitant infection is positive.

The authors would like to stress the fact that close follow-up with specialized monitoring of the velopharyngeal function when possible (physical examination ± nasometry) are mandatory in order to exclude a progressive condition. We propose a management algorithm based on this review's findings (Fig. 4): initially CNS imaging could be omitted if and only if after a complete clinical assessment the aforementioned criteria of "sudden-onset, isolated and unilateral" character are fulfilled. Another element that should be added is that a close follow-up, for example

1 week after the first evaluation and parental information on alarm signs are essential. If during follow-up new findings appear CNS imaging should be obtained immediately (dashed arrow in Fig. 4).

Examination of the velopharyngeal function is probably overlooked in everyday practice. Therefore, the exact incidence and prevalence of acquired VPI in general is difficult to estimate. Our literature review does not allow providing incidence/prevalence data but shows that reports on acute isolated VPI are scarce and probably understates the true scope of this problem. Whether this is due to a true low incidence or to low reporting rates remains an unanswered question.

Conclusions

Acute isolated VPI seems to be most often an idiopathic condition, but some cases could be associated with an infectious process. This diagnosis represents a subtype of VPI with a benign and most often self-limiting course, although incomplete recovery is seen in one-third of the cases. Complementary imaging studies do not seem to be initially necessary, but a close follow-up is mandatory to eliminate progressive underlying causes of VPI.

References

1. Conley SF, Gosain AK, Marks SM, Larson DL (1997) Identification and assessment of velopharyngeal inadequacy. Am J Otolaryngol 18(1):38–46
2. Seif S, Dellon AL (1978) Anatomic relationships between the human levator and tensor veli palatini and the eustachian tube. Cleft Palate J 15(4):329–336
3. Rouillon I, Leboulanger N, Roger G, Marlin S, Garabédian EN (2009) L'insuffisance vélopharyngée chez l'enfant. Ann Otolaryngol Chir Cervicofac 126(3):155–161
4. Ruda JM, Krakovitz P, Rose AS (2012) A review of the evaluation and management of velopharyngeal insufficiency in children. Otolaryngol Clin North Am 45(3):653–669
5. Auberge C, Ponsot G, Gayraud P, Bouygues D, Arthuis M (1979) Les hémiparalysies isolées et acquises chez l'enfant. Arch Fr Pediatr 36(3):283–286
6. Soares-Fernandes JP, Maré R (2006) Isolated velopalatine paralysis associated with parvovirus B19 infection. Arq Neuropsiquiatr 64(3-A):603–605
7. Nussey AM (1977) Paralysis of palate in a child. Br Med J 2(6080):165–166
8. Prasad PL, Prasad AN, Patnaik SK (2007) Unilateral palatal palsy with viral hepatitis. Indian J Pediatr 74(11):1039–1040
9. Sondhi V, Patnaik SK (2011) Isolated idiopathic unilateral paralysis of soft palate and pharynx. Indian Pediatr 48(3):237–239
10. Fleta Zaragozano J, Jiménez Vidal A, Meavilla Olivares S, Alonso Curcó X, Olivares López JL, Alfonso Collado I (2006) Hemiparálisis velopalatine idiopática. An Pediatr (Barc) 65(6):623–625
11. Alp H, Tan H, Altunkaynak S, Orbak Z (2005) Idiopathic unilateral paralysis of the palate in childhood. Pediatr Neurol 33(2):134–135
12. Jary A, Maillard L, Raffo E, Ducrocq X, Braun M, Vespignani H (2004) Paralysie vélopalatine isolée aiguë reversible de l'enfant. Rev Neurol 160(8–9):836–838
13. Villarejo-Galende A, Camacho-Salas A, Penas-Prado M, García-Ramos R, Mendoza MC Simón de las Heras R, Mateos-Beato F (2003) Parálisis aislada unilateral del velo del paladar: presentación de un caso y revisión de la literatura. Rev Neurol 36(4):337–339
14. Auvin S, Cuvellier JC, Vallée L (2003) Isolated recurrent palatal palsy in a child. Neuropediatrics 34(5):278–279
15. Mon Y, Nakamura N (2000) Velopharyngeal palsy: a case report. Rinsho Shinkeigaku (Clin Neurol) 40(4):364–366
16. Cuvellier JC, Cuisset JM, Nuyts JP, Vallée L (1998) Acquired and isolated asymmetrical palatal palsy. Neuropediatrics 29(6):324–325
17. Crovetto MA, Aguirre JM, Municio A, Perez-Rojo A, Saint-Gerons S (1988) Idiopathic paralysis of the palate in childhood. Br J Oral Maxillofac Surg 26(3):241–243
18. Roberton DM, Mellor DH (1982) Asymmetrical palatal paresis in childhood: a transient cranial mononeuropathy? Develop Med Child Neurol 24(6):842–846
19. Sullivan JL, Carlson CB (1976) Isolated temporary pharyngeal paralysis in childhood. Lancet 2(7990):863
20. Edin M, Sveger T, Tegner H, Tjernström O (1976) Isolated temporary pharyngeal paralysis in childhood. Lancet 1(7968):1047–1049
21. Shimokawa T, Yi SQ, Izumi A, Ru F, Akita K, Sato T, Tanaka S (2004) An anatomical study of the levator palatini and superior constrictor with special reference to their nerve supply. Surg Radiol Anat 26(2):100–105
22. Shimokawa T, Yi S, Tanaka S (2005) Nerve supply to the soft palate muscles with special reference to the distribution of the lesser palatine nerve. Cleft Palate Craniofac J 42(5):495–500
23. Lago PM, Guzmán MG, Sarmiento L, Pérez AB, Alvarez M, Capó V, Avalos I, Flores GK (2001) Mechanism of enterovirus involvement in epidemic neuropathy: hypothesis regarding pathophysiology. Med Hypotheses 56(3):339–347
24. Stanek G, Wormser GP, Gray J, Strle F (2012) Lyme borreliosis. Lancet 379(9814):461–473
25. Fahrer H, van der Linden SM, Sauvain MJ, Gern L, Zhioua E, Aeschlimann A (1991) The prevalence and incidence of clinical and asymptomatic Lyme borreliosis in a population at risk. J Infect Dis 163:305–310
26. Lefaivre JF, Cohen SR, Riski JE, Burstein FD (1997) Velopharyngeal incompetence as the presenting symptom of malignant brainstem tumor. Cleft Palate Craniofac J 34(2):154–158