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L. Denoix, P. Gatignol, N. Guevara, M. Montava, et al.

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French Society of ENT (SFORL) guidelines. Management of acute Bell's palsy

**Fioux M^a, Franco-Vidal V^b, Devic P^c, Bricaire F^d, Charpiot A^e, Darrouzet V^b, Denoix L^a,
Gatignol P^f, Guevara N^g, Montava M^h, Roch JAⁱ, Tankéré F^f, Tronche S^j, Veillon F^k,
Vergez S^l, Vincent C^m, Lamas G^f, Tringali S^{a,*}**

^a Service d'Otologie et d'Otoneurologie, Centre Hospitalier Lyon Sud, Hospices Civils de Lyon, Chemin du Grand Revoyet, 69495 Pierre-Bénite, France.

^b Service ORL et Chirurgie Cervico-Faciale, Hôpital Pellegrin, Place Amélie Raba-Léon, 33076 Bordeaux, France.

^c Service de Neurologie Clinique et Fonctionnelle, Centre Hospitalier Lyon Sud, Hospices Civils de Lyon, Chemin du Grand Revoyet, 69495 Pierre-Bénite, France.

^d Service des Maladies Infectieuses, Hôpital Pitié-Salpêtrière, AP-HP, 47-83 Boulevard de l'Hôpital, 75013 Paris, France

^e Service ORL et Chirurgie Cervico-Faciale, Hôpital de Hautepierre, 1 Avenue de Molière, 67200 Strasbourg, France

^f Service ORL et Chirurgie Cervico-Faciale, Hôpital Pitié-Salpêtrière, AP-HP, 47-83 Boulevard de l'Hôpital, 75013 Paris, France

^g Service ORL et Chirurgie Cervico-Faciale,

Centre Hospitalier de Nice, IUFC, 31 Avenue de Valombrose, 01600 Nice, France

^h Service ORL et Chirurgie Cervico-Faciale, Hôpital de la Conception, AP-HM, 147 Boulevard Baille, 13005 Marseille, France

ⁱ Service de Radiologie, Hôpital Privé Jean Mermoz, 55 Avenue Jean Mermoz, 69008 Lyon,
France

^j SFORL, 26 Rue Lalo, 75016 Paris, France

^k Service de Radiologie, Hôpital de Hautepierre, 1 Avenue de Molière, 67200 Strasbourg,
France

^l Service ORL et Chirurgie Cervico-Faciale, Hôpital Larrey, 24 Chemin de Pouvoirville,
31059 Toulouse, France

^m Service ORL et Chirurgie Cervico-Faciale, Hôpital Salengro, Rue Michel Polonowski,
59037 Lille, France

* Corresponding author:

E-mail: stephane.tringali@chu-lyon.fr (Stéphane Tringali)

Service d'Otologie et d'Otoneurologie, Hospices Civils Lyon, Centre Hospitalier Lyon Sud,
165 Chemin du Grand Revoyet, 69310 Pierre-Bénite, France

Tel. +33 4 72 66 64 15

Fax +33 4 78 86 33 36

Abstract

The authors present the guidelines of the French Society of ENT and Head and Neck Surgery (SFORL) regarding the management of Bell's palsy in adults. After a literature review by a multidisciplinary workgroup, guidelines were drawn up based on retrieved articles and group-members' experience, then read over by an independent group to edit the final version. Guidelines were graded A, B, C or "expert opinion" according to decreasing level of evidence.

Thorough ENT and neurological clinical examination is recommended in all patients presenting with peripheral facial palsy to confirm diagnosis of Bell's palsy. MRI with gadolinium enhancement should explore the entire course of the facial nerve, if possible within the first month. ENMG should be performed to assess prognosis for recovery. In confirmed Bell's palsy, corticosteroid therapy should be implemented as early as possible (ideally within 72 hours) at a dose of 1mg/kg/day for 7-10 days. Antiviral therapy should be associated to steroids in patients with severe and early-onset disease and in Ramsay-Hunt syndrome. Isolated antiviral therapy is not recommended. To date, there is no evidence that surgical facial nerve decompression provides benefit.

Keywords:

Bell's palsy; Drug therapy; Physical therapy; ENMG; Imaging

Introduction

Bells' palsy is the most frequent form of peripheral facial palsy, 8-12% of which, however, are of tumoral origin [1,2] (level of evidence, 4). Progression in Bell's palsy is spontaneously favorable, with more than 70% complete resolution within 6 months [3,4] (level of evidence, 3) and more than 80% subtotal resolution (House-Brackmann grade I or II) [5] (level of evidence, 3). Prognosis is thus usually good, but in a frequent pathology with severe social impact if recovery is incomplete. US and Canadian guidelines date respectively from 2013 and 2014 [6,7] (level of evidence, 1). The French Society of ENT and Head and Neck Surgery (SFORL) decided to update its own guidelines for Bell's palsy. The role of clinical examination, indications for complementary biological and radiological examinations, severity diagnosis, role of drug therapy, management of Ramsay-Hunt syndrome, management of ocular involvement, and the roles of rehabilitation, acupuncture, hyperbaric oxygen therapy and surgical decompression were discussed in drawing up these new guidelines.

The Medline and PubMed databases were searched for the period 1975-2019. Only French or English language articles were selected. Guidelines were graded A, B, C or "Expert opinion" according to decreasing level of evidence, in line with the French National Agency for Accreditation and Evaluation in Health (ANAES) guide to literature analysis and recommendation grading [8] (level of evidence, 1).

Guidelines:

1. Role of clinical examination

Bell's palsy is a form of peripheral facial palsy with sudden onset over 24-48 hours, involving the superior and inferior areas of the hemiface and affecting voluntary, autonomic and emotional motricity. It may be preceded by auricular or retroauricular pain, dysgeusia, hyperacusis and/or facial paresis. Diagnosis is by elimination. Clinical examination should comprise fine analysis of facial motricity in priority to confirm the peripheral nature of the palsy, with complete ENT and neurologic examination [5,9,10] (level of evidence, 2).

The peripheral nature of the palsy is based on:

- Balanced involvement in superior areas (reduction or abolition of forehead wrinkles and of eyelid closure (Charles Bell sign) and/or of the nasogenial groove) and inferior areas (lowered labial commissure and/or impossibility of blowing out the cheeks);
- no autonomic/voluntary dissociation;
- normal neurologic examination (somatosensory and motor central pathways and other cranial nerves).

- Confirmation of the peripheral nature of the facial palsy. Grade B
- In patients presenting for peripheral facial palsy, complete clinical neurologic and ENT examination should be performed, with otoscopy and parotid and cervical palpation. Involvement of the superior and inferior facial areas and absence of autonomic-voluntary dissociation should be screened to confirm peripheral status. Neurologic examination can rule out involvement of the somatosensory and motor central pathways and other cranial nerves. Grade B

Palsy progression parameters are essential to etiologic diagnosis. Progression of Bell's palsy is rapid, unlike other etiologies, and notably the tumoral etiologies that comprise the majority of differential diagnoses [11] (level of evidence, 2). The 2013 American Academy of Otolaryngology – Head and Neck Surgery Foundation guidelines set a threshold of 72 hours [6] (level of evidence, 1). Although Bell's palsy shows recurrence in 6% of cases [9] (level of evidence, 2), prior history of facial palsy casts doubt on a diagnosis of Bell's palsy [12] (level of evidence, 4).

- In case of peripheral facial palsy progressing beyond 72 hours after onset or showing fluctuation or recurrence or bilateral involvement, diagnosis of Bell's palsy should be questioned and tumoral causes screened for. Grade A
- In case of peripheral facial palsy even with rapid progression but associated with abnormal otoscopy or parotid or cervical lymph node palpation or ipsilateral hearing loss, dizziness or other neurological signs, diagnosis of Bell's palsy should be questioned. Grade B

There is no reason why isolated facial nerve injury should be associated with hearing loss; even so, audiometry seems mandatory to screen for an ipsilateral conductive component, suggesting intratemporal facial nerve tumor, or a sensorineural component, suggesting extrinsic facial nerve compression in the pontocerebellar angle. Tympanometry, performed in the same step, screens for stapedial reflex, which be useful for prognosis and localization [13,14] (level of evidence, 4)

- Audiometric assessment should be systematic in case of Bell's palsy. Expert opinion.
- Stapedial reflex screening should be systematic in case of Bell's palsy for prognostic purposes. In case of persistent stapedial reflex in severe peripheral facial palsy, a

diagnosis of Bell's palsy should always be questioned and extra-temporal cervical or parotid lesions should be screened for. Expert opinion.

Estimating neural involvement is essential to initial management of Bell's palsy, to grade severity and monitor progression. The most widely used classification in the initial phase is still that of House and Brackmann (HB) [15]. The Sunnybrook classification provides better sequela assessment and is more useful in follow-up.

- In case of Bell's palsy, initial work-up should assess facial involvement severity on a standardized grading system (House-Brackmann classification) for inclusion in the medical file. Expert opinion.

2. Laboratory testing

The 2013 US and 2014 Canadian guidelines [6,7] (level of evidence, 1) did not recommend any biological examinations in Bell's palsy. Even so, glycated hemoglobin (HbA1c fraction) assay may be useful in case of known diabetes, to control balance and prevent decompensation under high-dose corticosteroids [16] (level of evidence, 4). Likewise, complete blood count can screen for increased neutrophil/lymphocyte ratio, which is of poor prognosis [17] (level of evidence, 4).

- Hemogram (complete blood count) and fasting glycemia should be assessed, possibly completed by glycated hemoglobin (HbA1c fraction) assay in diabetic patients with Bell's palsy. Expert opinion.

Life-style factors may indicate HIV serology: HIV infection may induce peripheral facial palsy at any time, but especially at primo-infection [18] (level of evidence, 4). Serologic screening for Lyme disease is recommended, as incidence is increasing, at 6-20% of peripheral facial palsies depending on the geographical region [1,19] (level of evidence, 4).

- Serologic screening for Lyme disease should be systematic in Bell's palsy. HIV, VZV and HSV serology may also be performed, depending on the context. Expert opinion.

3. Role of imaging

The latest US and Canadian guidelines [6,7] (level of evidence, 4) do not consider imaging to be contributive in Bell's palsy when clinical and audiometric assessment is normal. However, recent data [1,2] (level of evidence, 4) show that 8-12% of cases of suspected Bell's palsy are of tumoral origin, including 30% malignancies [2] (level of evidence, 4). We therefore consider it essential to rule out differential diagnosis on adapted imaging within a month of symptom onset. MRI is the examination of choice, being alone in exploring the entire neural course. It should include brain and parotid, with gadolinium enhancement. In Bell's palsy, there is characteristic but non-specific enhancement of the facial nerve ipsilateral to the neurologic lesion [20–22] (level of evidence, 4). Emergency imaging, on the other hand, is not necessary if symptomatology is typical of Bell's palsy [23] (level of evidence, 4).

- In suspected Bell's palsy, contrast-enhanced MRI should be used to study the course of the facial nerve, including the parotid portion, within 1 month if possible, to contribute to positive diagnosis and rule out tumoral etiology.
- MRI is an emergency examination only in case of atypical clinical presentation.
- It should be borne in mind that diffusion-weighted MRI to rule out stroke does not explore the facial nerve and is thus insufficient for work-up in Bell's palsy.
- In the absence of the usual favorable progression, imaging should be repeated at 6 months.
- Brain or temporal CT is not indicated in work-up for Bell's palsy.

Expert opinion.

4. Diagnosis of severity

Electroneuromyography (ENMG) is fundamental for assessing the clinical recovery profile, but should not be performed too early [24] (level of evidence, 4), else it would underestimate compound motor action potential (CMAP) due to the chronology of axon degeneration. Also, absence of motor unit potential (MUP) on effort has no great prognostic value until day 15. [25] (level of evidence, 4).

- ENMG should not be performed before day 8 to assess prognosis in Bell's palsy, given the poor prognostic value of the results, which are moreover poorly reproducible. Expert opinion.

According to a national expert consensus and the literature data, ENMG can classify prognosis of recovery according to: i) myelinic involvement, with good prognosis; ii) mixed involvement, with intermediate prognosis; and iii) severe axonal involvement, with poor prognosis [26] (level of evidence, 4).

- Electroneuromyography should be performed in severe facial palsy (HB grade V or VI) between days 9 and 20 to assess recovery prognosis. Expert opinion.
- For optimal, prognostic value, ENMG should include:
 - Study of bilateral facial nerve motor conduction recorded in several muscles including at least 1 peri-oral muscle (nasolabial groove, orbicularis oris, mentalis). Ideally, double recording simultaneously studies muscles with superior (nasalis, nasolabial groove) and inferior branch innervation (orbicularis oris, on the midline method). The nasolabial groove muscles may be more relevant for the patient, being preponderant in smiling. The prognostic value of CMAP amplitude in the orbicularis oculi seems to be controversial.
 - Study of blinking reflex.
 - Study of myography in >2 muscles, from the superior and inferior facial nerve branches, screening for spontaneous activity at rest and assessing recruitment of motor unit under effort. Expert opinion.

5. Role of medical treatment

Medical treatment for Bell's palsy has been debated for many years, as progression is spontaneously favorable, with more than 70% complete recovery in 6 months [3,4] (level of evidence, 3) and more than 80% subtotal recovery (HB grade I or II) [5] (level of evidence, 3). Recovery rate correlates with initial severity, ranging from 61% in total palsy to 94% in partial palsy [5] (level of evidence, 3). Most authors agree that corticosteroids are effective [27,28] (level of evidence, 1), as confirmed by US and Canadian guidelines [6,7] (level of evidence, 1).

- In Bell's palsy, corticotherapy with prednisolone or methylprednisolone should be implemented as early as possible (ideally, within 72 hours). Grade A.

Many studies have focused on the form and dosage of such corticotherapy, but only one robust randomized study compared intravenous versus oral route and found no difference in recovery [29] (level of evidence, 2). Intravenous administration seemed to give better results, but in small comparative studies vitiated by biases [30] (level of evidence, 4).

- There are at present no level 1 or 2 studies comparing dosages to support any high-grade recommendations.
- Bell's palsy should be treated by corticosteroids at 1mg/kg/day for 7-10 days. Expert opinion.
- In severe forms (HB grade V or VI), 10 days' high-dose corticotherapy (2mg/kg/day) seem justified unless contraindicated. Expert opinion.

Studies of transtympanic administration are insufficient, with low level of evidence.

- Isolated transtympanic administration is not recommended for Bell's palsy. Expert opinion.

All multicenter randomized double-blind placebo-controlled studies since 2007 confirmed the absence of significant improvement in efficacy with isolated treatment by aciclovir or valaciclovir [31–33] (level of evidence, 1).

- Isolated antiviral treatment is not recommended in Bell's palsy. Grade A

US and Canadian guidelines suggest associating antiviral treatment to corticotherapy in the initial phase of Bell's palsy [6,7] (level of evidence, 2). Two recent meta-analyses [34,35] (level of evidence, 2) and a 2019 update [36] (level of evidence, 2) confirmed the interest of this bi-therapy, with absence of antiviral side-effects and possible limitation of sequelae.

- In severe Bell's palsy treated early (within 72 hours), antiviral treatment should be associated to corticosteroids. Expert opinion.

6. Medical treatment of Ramsay-Hunt syndrome

Ramsay-Hunt syndrome resembles Bell's palsy, with some clinical differences and much greater severity. The most important difference is its tendency to recurrence [11] (level of evidence, 3). Progression is more severe, with 50-85% of patients showing sequelae [11,37,38] (level of evidence, 3).

- In Ramsay-Hunt syndrome, 7 days' corticotherapy with prednisolone or methylprednisolone associated to antiviral treatment should be initiated as early as possible. Grade B

7. Eye-care in Bell's palsy and Ramsay-Hunt syndrome

The main functional complications in Bell's palsy are ophthalmologic: keratitis, corneal ulcer, panophthalmia. They can be prevented by systematic administration of eye-drops, protective gel or artificial tears several times daily and especially at night [6,27,39–41] (level of evidence, 4). No randomized studies have compared results with or without such ophthalmic treatment, but failure to administer systematic treatment leads to potentially catastrophic complications.

- Ophthalmologic treatment comprising local care, nocturnal occlusion and patient education should be systematic and as early as possible. In case of painful red eye, treatment should be entrusted to an ophthalmologist. Ophthalmologic monitoring should continue for several weeks once recovery has begun. Expert opinion.

8. Role of physical therapy in Bell's palsy

Physical therapy is classically applied in Bell's palsy persisting beyond the first months, to hasten recovery and limit sequelae. A recent meta-analysis (level of evidence, 4) of 12 studies for 872 patients reported high risk of bias due to variations in techniques and timing [42] (level of evidence, 4). However, it is clear that facial muscle rehabilitation improves facial function when Bell's palsy fails to resolve [6,43,44] (level of evidence, 4).

- Treatment by a rehabilitation specialist (speech therapist or physiotherapist, with relevant qualifications) is recommended in severe Bell's palsy or in case of factors for poor recovery. Expert opinion.

Since 1984, electrotherapy has been considered deeply harmful, shortening the muscle fibers and hindering recovery [45] (level of evidence, 4).

- Electrotherapy and forced exercise (chewing gum, biofeedback) are formally contraindicated in Bell's palsy. Expert opinion.

Acupuncture was reported to be superior to drug treatment in a Chinese meta-analysis, but this finding is dubious due to considerable bias induced by diversity of techniques [46] (level of evidence, 4). Evidence is lacking for isolated acupuncture according to the most recent US guidelines [6] (level of evidence, 4).

- Acupuncture is not recommended in Bell's palsy.
- As few studies had sufficient level of evidence, acupuncture cannot be recommended in acute Bell's palsy. Grade C

9. Role of hyperbaric oxygen therapy in Bell's palsy

The principle of hyperbaric oxygen therapy is to spread oxygen within tissue and thus reduce facial nerve hypoxia, enhancing recovery. A 2012 meta-analysis found that all studies had low levels of evidence, with serious biases [47] (level of evidence, 4).

- Hyperbaric oxygen therapy is not recommended in Bell's palsy. Grade C

10. Role of follow-up in Bell's palsy

Clinical and ophthalmological follow-up should be continued for several months after recovery begins, to ensure against complications, and ophthalmic complications in particular [6,7] (level of evidence, 4).

- When Bell's palsy does not fully resolve, prolonged surveillance enables treatment to be adapted to functional progression. Grade C

11. Surgical facial nerve decompression in acute Bell's palsy

The benefit of surgery in Bell's palsy is at present unproven. The benefit of decompression is difficult to demonstrate as many factors can affect outcome: patient selection, timing of surgery, and method and approach. However, although efficacy cannot be proved, we consider that surgery should still be proposed as an alternative treatment, as advocated by Lee et al. in their 2019 meta-analysis [48] (level of evidence, 4).

- There is at present no proof that facial nerve decompression provides benefit in acute Bell's palsy. Even so, given the diversity of results and techniques, we believe that further well-conducted studies should be performed before declaring such treatment ineffective. Expert opinion.

Diagnosis of Bell's palsy should be established with certainty before considering surgical decompression. Diagnosis is founded on clinical, electromyographic and radiological criteria. There seems to be consensus as to which part of the nerve is to be decompressed in the light of radiological and surgical findings.

- If surgical facial nerve decompression is implemented, it must include the meatal foramen, the labyrinthine segment, the beginning of the second part of the nerve, and the geniculate ganglion. A transmastoid or a supratemporal approach may be used, but the latter is the gold-standard.
- There is no benefit in isolated decompression of the third part of the facial nerve in Bell's palsy. Expert opinion.

In severe forms, decompression should be performed early to avoid irreversible nerve injury [49,50]. If ENMG shows >90% degeneration, decompression should be performed rapidly: later surgery shows progression comparable to standard medical treatment alone [50] (level of evidence, 2).

- Any facial nerve decompression should be performed within 30 days and ideally within 14 days of electrically proven onset. Expert opinion.

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