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Bell's Palsy: The Spontaneous Course of 2,500 Peripheral Facial Nerve Palsies of Different Etiologies

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Objective—The Copenhagen Facial Nerve Study aims to explain the spontaneous course of idiopathic peripheral facial nerve palsy which occurs without any kind of treatment. In this study Bell's palsy and idiopathic palsy are considered to be synonymous and specify an acute, monosymptomatic, unilateral peripheral facial paresis of unknown etiology.

Material and methods—The material includes 2,570 cases of peripheral facial nerve palsy studied during a period of 25 years. It includes 1,701 cases of Bell's palsy and 869 of non-Bell's palsy. In the total patient sample, 116 had herpes zoster, 76 were diabetic, 46 were pregnant and 169 were neonates. A total of 38 different etiologies were observed. At the first consultation a standard ENT examination was performed, including a thorough description of the grade and localization of the paresis, taste, stapedius reflex and nasolacrimal reflex tests and acoustic-vestibular examination. Follow-up was done once a week during the first month and subsequently once a month until normal function was restored or for up to 1 year.

Results—The initial examination revealed 30% incomplete and 70% complete palsies. Follow-up showed that in 85% of patients function was returned within 3 weeks and in the remaining 15% after 3–5 months. In 71% of patients normal mimical function was obtained. Sequelae were slight in 12% of patients, mild in 13% and severe in 4%. Contracture and associated movements were found in 17% and 16% of patients, respectively.

Conclusion—A survey of the literature showed that no kind of treatment, including prednisone, was able to give a better prognosis. The use of prednisone raises a big ethical problem because no evidence of its efficacy exists and the euphoric side-effect induces a false feeling of benefit in the patients. *Key words*: *Bell's palsy*, *facial nerve*, *idiopathic facial nerve paresis*, *natural history*, *spontaneous course*.

INTRODUCTION

The facial expressions of human beings fascinate me because they convey both the lowest, most bestial pleasures and the strongest and gentlest emotions of the spirit. With these words, Charles Bell defined the importance of peripheral facial paralysis, which eliminates facial symmetry, one of the attributes of beauty, and thus creates a disfigured and distorted face (1).

Facial nerve paralysis has been known since ancient times by the Egyptians, Greeks, Romans, Incas and other native cultures (2, 3). The oldest artistic representation of facial nerve paralysis is a clay head from Egypt, $\approx$4,000 years old, showing a right peripheral facial nerve paralysis. A small painted earthenware statuette of a woman with a left-sided facial paralysis from Crete, Greece, has been dated $\approx$7th to 6th century BC. A vase found in Carthage and dated $\approx$250 BC (from the third Punic war 249–246 BC) depicts a peripheral facial nerve palsy on the left side. Mochica ceramics from Peru (AD 200–700) made in clay and carefully painted represent typical asymmetrical faces caused by facial nerve paralysis.

In the Middle Ages and during the Renaissance many artists portrayed figures with asymmetrical and distorted faces. The portrait of Mona Lisa is the most famous and her enigmatic smile has been discussed for many years all over the world and at several facial nerve symposiums. There is a weakness of the corner of the mouth, but on which side? She cannot be asked to grimace to reveal which side was affected. Leonardo da Vinci (1452–1519) took 4 years to paint Mona Lisa but the painting was never finished (4). The expression of a face depends to a large extent (5) on the corners of the eyes and mouth but Leonardo blurred these parts of the face very carefully, so that it is very difficult to guess her mood (6). Furthermore, the painting is asymmetrical (there is a lower left horizon), which gives the left side of the face a leaner and more erect impression (6). Attempts to interpret the smile of Mona Lisa have been numerous. A guess is that it could be the smile of Leonardo's mother, who died before he was 5 years old (4, 7, 8). The smile of Mona Lisa was used by Leonardo in some of the paintings he made after his masterpiece and is called "Leonardesque" (4). The conclusion is that, from the artist's point of view, Mona Lisa did not suffer from a peripheral facial nerve palsy, as has been misdiagnosed by medical doctors (9).

The first medical studies of the disease should be attributed to Avicenna (10), who was the first to record the differences between central and peripheral facial paralysis. If the disease that produces paralysis comes from the middle of the brain, one half of the

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body is paralyzed. If the disease is not in the brain but instead in the nerve, then only what depends on this nerve is paralyzed.

The representation of facial nerve palsy in medical publications began in the 18th century. In 1797, Professor Niclaus A. Friedreich from Würzburg, Germany treated three patients with idiopathic facial nerve paresis and documented recovery of normal function (11). The observation was published in the German medical literature in 1798 under the title "Paralysis Musculorum Faciei Rheumatica". The first English review appeared in 1800 in the *Annals of Medicine* published in Edinburgh and it is possible that Charles Bell, who was studying medicine in Edinburgh at the time, read this paper. Bell (later Sir Charles Bell) described the innervation of the facial muscles and the skin of the face and consequently the trigeminal nerve is called Bell's nerve. Eventually the eponym "Bell's palsy" became synonymous with idiopathic peripheral facial nerve paralysis. However, Friedreich described the syndrome 23 years before Bell.

During the 19th century the treatment of peripheral facial nerve palsies was anti-rheumatic. At the beginning of the 20th century electrical stimulation was used and some very ingenious apparatuses were constructed. In 1932 Balance and Duel (12) published the results of inserting a free nerve graft between the cut ends of the facial nerve and also advocated decompression of the mastoid segment of the facial nerve for Bell's palsy when its degeneration delayed recovery. This was the beginning of the modern era of facial nerve surgery.

During the following three decades, the number of facial nerve decompression operations increased rapidly. The leading experts were Cawthorne from London (13), Miehlke from Germany (14), Jongkees from The Netherlands (15), Fisch from Switzerland (16) and Kettel from Denmark (17), who was the organizer of the First International Symposium on Facial Nerve Surgery held in Copenhagen in 1964 (18). At that time, the surgeon's view was that peripheral facial paralysis should be treated with decompression after 2 months in cases of continued paralysis. The discussions at the symposium revealed a great deal of skepticism, especially from neurologists and neurophysiologists, regarding the efficacy of the decompression operations. Sunderland (19, 20) described nerve injuries based on the pathology of the nerve trunk and classified five degrees of damage (Table I). Furthermore, he combined the severity of the injuries with the time of recovery and was the first to create a profile of the recovery of a motoric nerve palsy.

AIM OF THE INVESTIGATION

In Copenhagen it was decided to study the natural history of Bell's palsy. The aim of this "Copenhagen Facial Nerve Study" was to provide a description of the spontaneous course of idiopathic peripheral facial palsy, i.e. that which occurs without any kind of treatment. The prospective study was designed to include a large number of patients, with no exclusion of special groups of patients, and to involve adequate follow-up, exact descriptions of sequelae and a statistical analysis with significant conclusions. It was decided not to publish the results of the investigation until the conclusions were statistically significant.

Preliminary observations were presented in Zürich in 1976, Pittsburgh in 1977, Los Angeles in 1981, Paris and Rio de Janeiro in 1988, Cologne in 1992, Matsuyama in 1997 and Berlin in 2000.

ETIOLOGY

Theories and hypotheses

The aim of this investigation is not to list or discuss all possible hypotheses or theories but to describe the spontaneous course of Bell's palsy. Nevertheless, a short summary of these theories is included. For reviews, see Kettel (17), Miehlke (14) and May (21). Friedreich (11) hypothesized that the cause of facial paralysis in his three patients was "rheumatic" because of exposure to cold often followed by fever, chills and local pain and swelling in and around the neck. Brunninghausen speculated that the paralysis arose from the nerve sheath becoming thickened and compressed in the stylomastoid foramen. Berard in 1836 repeated this hypothesis. The cold hypothesis, or paralysis e frigore, maintained that exposure to draughts produced the palsy. The ischemic hypothesis maintained that ischemia resulting from disturbed circulation in the vasa nervorum led to nerve injury. Later on a combination with secondary ischemia was advocated by a number of surgeons (13–17). The immunological hypothesis was introduced by McGovern and co-workers (22, 23).

During the last 25 years or more, viral infections have been proposed as causes of Bell's palsy. In 1972,

Table I. *Classification of nerve injuries after Sunderland (19, 20)*

Degree	Pathology	Recovery
1	Neuropraxia	Complete
2	Axonotmesis	Complete
3	Neurotmesis	Incomplete
4	Perineurium disruption	Non-functional
5	Complete disruption	None

Table II. *Timetable for examinations*

First examination as soon as possible
Once a week until function returned
Thereafter every second week
After 6 months once a month
Follow-up discontinued after function restored or after 1 year

McCormick (24) published his hypothesis suggesting that reactivation of herpes simplex virus type 1 (HSV-1) causes inflammation and edema in the bony fallopian canal and results in peripheral facial palsy. Adour et al. (25) accepted this theory. In contrast, Mulkens et al. (26) were unable to demonstrate a direct link with HSV-1. However, new molecular biological techniques, such as polymerase chain reaction (PCR), have proved to be more sensitive. Murakami et al. (27) studied 14 Bell's palsy patients in vivo using HSV DNA PCR. Endoneurinal fluid from the facial nerve and biopsies from the posterior auricular muscle were tested using PCR for the presence of HSV DNA. HSV DNA was detected in samples from 11/14 patients. Further experiments in the future will hopefully resolve this issue.

Definition

The term Bell's palsy is accepted in many Anglo-American countries to describe a peripheral paresis of the facial nerve, independent of etiology. Idiopathic peripheral facial nerve paresis is an acute, monosymptomatic, peripheral facial nerve paresis of unknown etiology. Formerly the disease was described as rheumatic or ischemic, as mentioned above. Patients suffering from an underlying disease or condition, for example collagenosis, pregnancy or diabetes mellitus, sometimes develop peripheral facial nerve palsies, but these pareses are never classified as Bell's palsy. However, a reduction in future in the group of idiopathic pareses is anticipated on account of better knowledge of many diseases and of the pathological conditions governing peripheral paresis. Bell's palsy and idiopathic peripheral facial nerve paresis are considered as synonymous in this study.

MATERIAL AND METHODS

Before the start of this investigation a lot of time was taken to work out a very specific plan for the study. As a result of 6 months of pilot studies it was possible to construct a timetable for the following years. The study design was as follows.

The patient was first examined as soon as possible after the onset of palsy, i.e. within 1–5 days after the onset of paresis (Table II). Follow-up examinations were performed once a week until the returning of function was observed. After 6 months, examinations were conducted once a month. Follow-up was discontinued after restoration of function or after 1 year. If a patient did not attend for examination a new appointment was mailed to them. If the patient still failed to attend, a telephone interview was conducted or the patient was asked to complete a questionnaire. All the patients lived within a radius of 25 km of the hospital and using the Danish Central Personal Register it was easy to trace them. The majority of patients failed to attend the follow-up examinations because normal function had been restored. Follow-up included 98% of all patients. Great importance was attached to the date of onset of palsy as well as the date of the first sign of returning of function. The questionnaire administered to the patients is shown in Table III. It should be stressed that patients suffering from acute peripheral facial palsy were always able to give the exact date and hour of the first sign of paresis.

Table III. *The questionnaire administered to the patients concerning history of Bell's palsy*

Beginning—palsy—date
Remission date—unchanged—regression
Facial palsy before—relatives with facial palsy
Head trauma—systemic disease—diabetes mellitus
Pregnancy—infections—skin eruptions
Colds—exposure to draught
Other cranial nerve symptoms
Headache—paresthesia—paresis
Otitis—hearing loss—tinnitus—vertigo
Postauricular pains
Before—simultaneous—after
Taste—phonophobia—tearflow—dry eye

The examination of the patient is very important and an exact description of the paresis must be made at the first examination (Table IV). Is the paresis complete or incomplete? Does it affect all branches or is it localized to only one or maybe two branches? Bell's palsy usually involves all branches, although not to the same degree. Patients with involvement of only one or perhaps two branches should arouse

Table IV. *Description of the standard examination of patients with Bell's palsy*

Routine ENT examination
Description of facial nerve palsy
Grade and localization
Cranial nerve check
Taste, stapedius reflex and nasolacrimal reflex tests
Acoustic and vestibular function tests
Laboratory tests

suspicion of another etiology, for example a parotid gland tumor.

All topographical tests were performed during the patient's first visit. These involved examination of taste, stapedius reflex and nasolacrimal reflex. These tests were repeated at each of the two subsequent visits, if the result of the preceding investigation was not normal. Patients also underwent acoustic and vestibular tests including tympanometry, audiometry, electronystagmography for spontaneous and positional nystagmus and a caloric test according to Hallpike.

If the anamnesis and objective findings indicated Bell's palsy, only a few laboratory tests were carried out; these always included measurement of blood pressure and a urine test for glucose. All patients were investigated for serum antibodies against HSV, herpes zoster and borreliosis. In patients with a second episode of palsy or those in whom function was not restored within 4 months an extensive battery of tests was performed, including cerebrospinal fluid (CSF) cell counting, differential cell counting, determination of Borrelia burghdorferi antibodies, CT and MRI scans and a variety of other tests.

Electrodiagnosis of the facial nerve

Duchenne in 1855 (28) was the first to use contraction of the facial muscles evoked by electrical stimuli to the nerve as a way of predicting recovery. In contrast to many lesions of peripheral nerves, electrodiagnosis of cranial nerves is hampered by one major difficulty, namely that in the majority of cases it is not possible to stimulate proximally to the site of the lesion. Thus, it is not possible to compare the response to proximal stimulation with the response to stimulation distally from the site of the lesion.

A very simple and widely-used electrical test is the so-called nerve excitability test (NET), in which the threshold of the electrical stimulus producing visible muscle twitch is determined. Although the NET is a very simple test, it has limitations and its accuracy is dubious. When using a stronger stimulation to obtain maximal contraction of the facial muscles the test is known as the maximal stimulation test (MST). According to Blumenthal and May (29), the NET was not reliable when the response was normal, as 42% of their patients with a normal response had residual facial function deficits 6 months later. Furthermore, Groves and Gibson (30) and Laumans (31) noted that some of their patients with an abnormal response to the NET recovered full function. To the best of my knowledge, the NET is not reliable or useful for predicting recovery from peripheral facial nerve palsies. Langworth and Taverner (32) recommended conduction velocity as the best parameter for predicting prognosis.

Electromyography is not very reliable at revealing recent facial nerve palsies and has no prognostic significance in Bell's palsy according to Buchthal (33). However, after $\approx 10-12$ days the so-called denervation potentials can be recorded and the test is also able to demonstrate regeneration.

Electrical tests are not routinely used in Bell's palsy patients in my department. The Department of Clinical Neurophysiology performed all the electrodiagnostic tests of the facial nerve in this study. The majority of cases were examined by Olsen (34), who in 1975 described the technique of electroneurography (ENoG). The method is identical to that used by Esslen in 1977 (35). ENoG is very useful for demonstrating degeneration. The principle is to compare the evoked potentials on the paretic side with those on the healthy side. For further information, see Olsen (34) and Esslen (35).

Statistical analyses

The statistical data analyses were performed by Arne Nørby Rasmussen, BScEE and Poul Aabo Osterhammel, EE, EDA. The χ^2 test was used to compare data between different groups of patients. $p = 0.05$ was considered significant. In addition, the Shapiro–Wilk test for normality was used with a significance level of $p = 0.05$.

Patients

The patients in this study came from the Copenhagen area during a 25-year period. The investigation included 2570 patients suffering from peripheral facial nerve paresis (Table V). There were 1,701 patients with Bell's palsy (66%) and 869 with non-Bell's palsy (34%).

RESULTS

Incidence of Bell's palsy

In this study "incidence" is defined as the number of cases per year in a population of 100,000 inhabitants. Studies in the literature have shown great variations in incidence. However, many of these studies cannot be considered as representative, because basic criteria have not been fulfilled. There must be a well-defined area and all patients should be included. The number of all incoming and outgoing inhabitants should be registered, as well as the age and sex of the population; however, only a few investigations meet these requirements. In this study the incidence of Bell's palsy was 32.

Recurring palsies and familial Bell's palsy

A total of 6.8% of Bell's palsy patients in the sample had previously suffered from facial palsy on either

Table V. *Etiologies of the 2,570 study patients*

Etiology	n
Idiopathic palsy	1,701
Neonatal age	169
Herpes zoster	116
Trauma	95
Diabetes mellitus	76
Pregnancy	46
Polyneuritis	44
Parotid tumor	43
Vascular (brainstem)	34
Hemifacial spasm	27
Sarcoidosis	21
Multiple sclerosis	20
Melkersson–Rosenthal syndrome	19
Collagenosis	18
Cholesteatoma of the middle ear	18
Children with bilateral palsy	18
Breast cancer with metastasis	16
Chronic otitis	15
Infectious mononucleosis	9
Leukemia	6
Malignant lymphoma	6
Cholesteatoma of the inner ear	5
Cancer of the middle ear	5
Borreliosis	4
AIDS	4
Neurinoma of the VIIth extracranial nerve	3
Bronchial cancer with metastasis	3
Nephropathia	2
Non-definite paresis	2
Hypothyreosis	2
Pemphigus	1
Granuloma eosinophilia	1
Tuberculosis	1
Poliomyelitis	1
Smallpox vaccine sequelae	1
Herxheimer reaction	1
Paget's disease	1
Hysteria	1

the same or the opposite site of the face. Some authors use the term "recurrent" palsy exclusively to describe paresis occurring on the same side of the face; paresis on the opposite side is termed "alternating". Although this classification is logical, the term "recurrent" is in general use and is therefore used in this study to describe both ipsi- and contralateral palsies. The occurrence of familial Bell's palsy is well known. In this study, 4.1% of all cases of Bell's palsy observed represented familial Bell's palsy.

Seasonal variation and clustering

The seasonal incidence of Bell's palsy has been discussed for many years. The explanation of findings of seasonal variation or clustering in some samples could be that the patient sample was too small. In this study the mean number of patients per month with Bell's palsy was 142 (range 126–156) (Fig. 1).

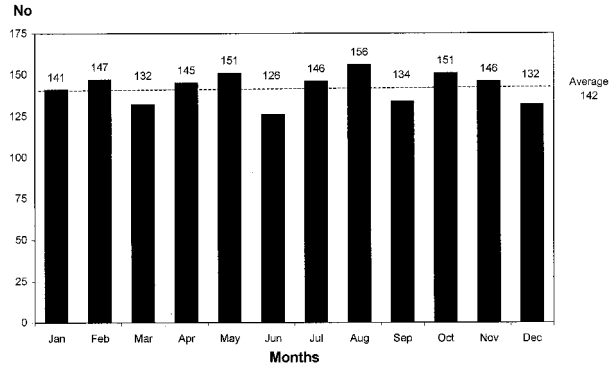

Fig. 1. Number of patients with Bell's palsy per month over the 25-year period of the study.

The Shapiro–Wilk test for normality revealed no significant difference from month to month ($p = 0.1$) and thus no seasonal variation or clustering was found.

Decade variation

Variations from year to year or from decade to decade could not be demonstrated because the number of patients with Bell's palsy has largely remained constant over the study period: mean 68 (range 48–89) (Fig. 2). The Shapiro–Wilk test showed no significant difference from year to year ($p = 0.4$).

Sex distribution

The question of a gender predominance among patients afflicted with idiopathic facial palsy has been discussed in the literature. One of the main reasons for the discussion has undoubtedly been the small amount of material available. This study included 1,701 patients with Bell's palsy, 818 (48.1%) of whom were male and 883 (51.9%) female. The underlying population comprises 47.8% males and 52.2% females, clearly indicating that there is no difference in sex distribution among patients with Bell's palsy ($0.8 < p < 0.9$).

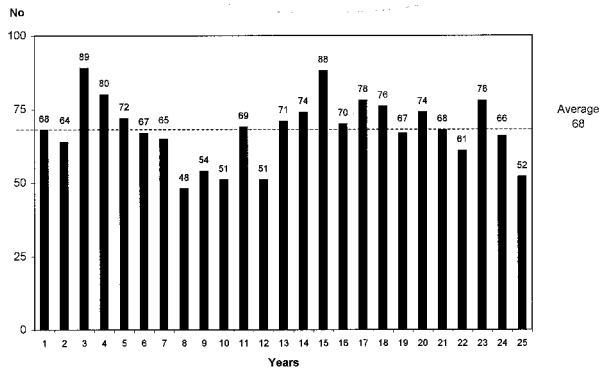

Fig. 2. Number of patients with Bell's palsy per year over the 25-year period of the study.

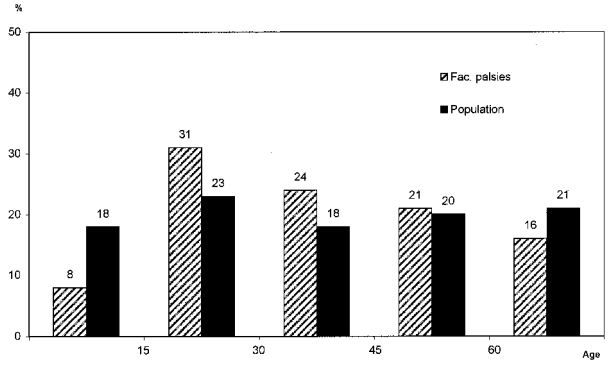

Fig. 3. Age distribution of patients with Bell's palsy in comparison with that of the underlying population.

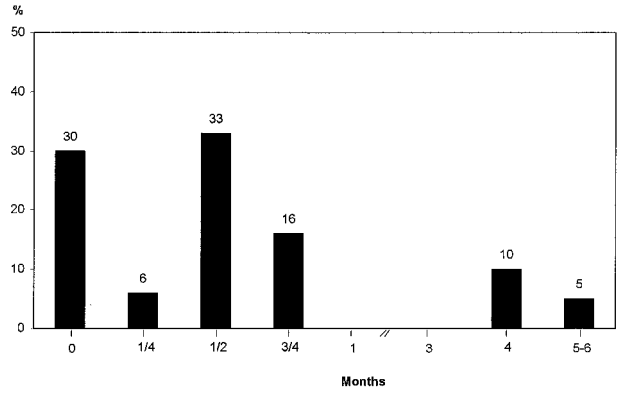

Fig. 4. Distribution of time of beginning recovery after the onset of paresis.

Side of the face

There was no difference in localization, with 828 (48.7%) right- and 873 (51.3%) left-sided palsies ($0.1 < p < 0.2$).

Age distribution

This subject has also been debated, without definitive conclusions having been reached. Figure 3 demonstrates the age distribution of patients with Bell's palsy in comparison to the age distribution of the underlying population. The incidence of Bell's palsy reaches a maximum between the ages of 15 and 45 years and this differs highly significantly from the age distribution of the underlying population ($p < 0.001$). The disease is significantly less common below the age of 15 years and above the age of 60 years ($p < 0.001$). For the group aged 45–59 years the incidence of Bell's palsy did not differ significantly from the age distribution of the underlying population ($p = 0.4$). The influence of age on the incidence of Bell's palsy would therefore seem to be convincingly demonstrated.

Symptoms

The most alarming symptom of Bell's palsy is of course the paresis itself. Approximately 50% of patients believe that they have suffered a stroke, 25% fear an intracranial tumor and the remaining 25% have no clear conception of what is wrong, but are extremely anxious. The distribution of symptoms among the patient sample is shown in Table VI. Postauricular pains, which were experienced by almost half the patients, are of the utmost interest. These pains occurred simultaneously with the palsy in $\approx 50\%$ of patients, whilst in 25% they occurred 2 or 3 days before the onset of palsy. The remaining 25% of patients experienced pains after the onset of palsy. The pains are located deep in the mastoid region, usually persist for one to several weeks and require analgesia.

Table VI. *Distribution of symptoms of Bell's palsy among the patient sample*

Symptom	n	%
Taste disorders	580	34
Phonophobia	234	14
Tear flow	1137	67
Dry eye	69	4
Postauricular pains	881	52

Every third patient will complain about taste disorders but when examined objectively four out of five show a reduced sense of taste. This difference can be explained by the fact that patients can still use the normal side of the tongue to taste.

Only a few patients are able to perceive restricted stapedius reflex paresis, which may lead to phonophobia or diplaucusis. In comparison, two out of three patients complain about tear flow. However, this is not caused by hypersecretion but by the diminished function of the musculus orbicularis oculi, which prevents tears from being transported medially to the lacrimal sac.

Time of beginning recovery

Very little interest has been paid in the literature to the question of recovery time and therefore it was felt worthwhile to record the pattern of remission for the idiopathic palsies. The time of the first sign of muscular movement in relation to the onset of palsy was recorded.

Of a total of 1,701 patients, 1,189 suffered from complete paralysis (70%) and 512 from incomplete paralysis (30%). Recovery occurred within 3 weeks for 1,448 patients (85%) and within 3–5 months for the remaining 253 patients (15%).

The results are shown in Fig. 4. By definition, all patients with incomplete paresis have function back at time zero, i.e. at the onset of paresis. In the first

week 6% of patients achieved remission, in the second week 33% and in the third week 16%. No patients achieved remission between 3 weeks and 3–5 months after the onset of paresis. This is because patients who showed improvement in the first 3 weeks had only partial degeneration and blocking of nerve conduction whilst patients who showed improvement after 3–5 months had total degeneration. After 3–5 months 10% of patients experienced remission and after 5–6 months an additional 5% experienced remission. This investigation shows that all patients diagnosed with Bell's palsy achieve some degree of muscular function. However, this does not imply that all patients achieve normal function. The period between the first 3 weeks without remission and after the third month without remission is one characterized by "hibernation of the facial nerve". Although the nerve seems to be dead it is in fact still alive and in the process of repairing the damage.

Complete recovery

The next question is naturally how many patients will achieve complete remission? In this study, of a total of 1,701 patients, 1,202 (71%) achieved normal facial nerve function. The second question is how long does it take before patients achieve normal function?

Prospects are decidedly better for the group with some remission within the first 3 weeks (Table VII). Patients with the poorest prognosis are of course those with total degeneration and late return of function. This group does not regain normal mimical function. Normal function is regained as early as within 2 months for the majority (58%) of all patients (Fig. 5). The possibility of normalization is very small after 3 months, at which time 64% of patients have regained normal function, and beyond 6 months no patients regained normal mimical function (Fig. 6). As noted before (Table VII), the incomplete Bell's palsy patients have a very good prognosis for full recovery (481/512; 94%). Of the patients with complete Bell's palsy, 721/1,189 (61%) regained normal facial muscle function. The difference between the two groups in terms of the number of patients who

Table VII. *Distribution of patients with initial incomplete and complete paresis who make a full recovery from Bell's palsy*

Paresis with full recovery	Initial		Final	
	n	%	n	%
Incomplete	512	30	481	94
Complete	1189	70	721	61

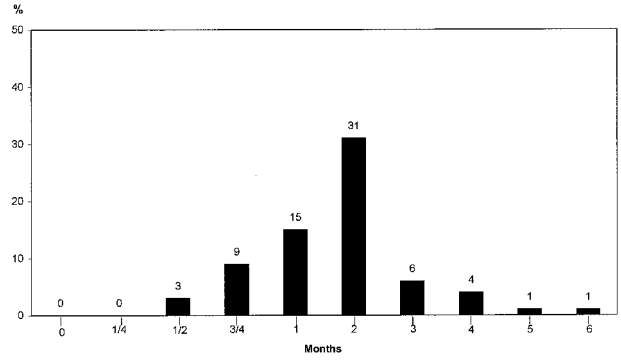

Fig. 5. Distribution of time of complete recovery after the onset of paresis.

achieved full recovery was highly significant ($p < 0.001$).

Gender and recovery

The numbers of males and females who experienced full recovery were 564 (69%) and 638 (72%), respectively. There was no statistically significant difference between the two groups ($p > 2$).

Factors influencing the final resuls

Time of beginning remission. The number of days between the onset of paresis and the beginning of remission is a very decisive factor in the degree of recovery (Fig. 7). A total of 94% of patients with incomplete paresis regained normal function. Of patients who showed remission in the first week, 88% regained normal function, as opposed to 83% of those who showed remission in the second week and 61% of those who showed remission in the third week. The prognosis for patients with incomplete paresis was significantly better than that for the group who recovered in the first week ($p = 0.03$). There was no significant difference in prognosis between patients who recovered in the first and second weeks ($p = 0.2$) but patients who recovered in the

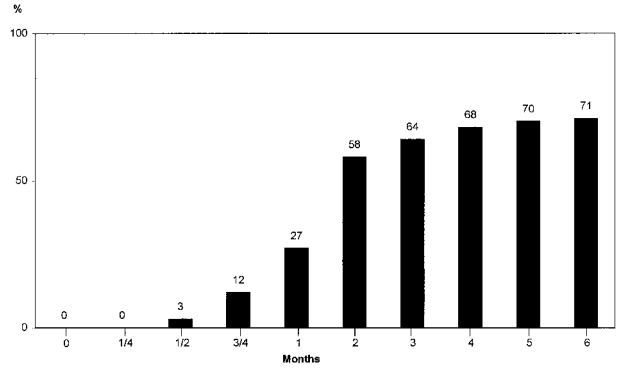

Fig. 6. Distribution of time of complete recovery (cumulative) after the onset of paresis.

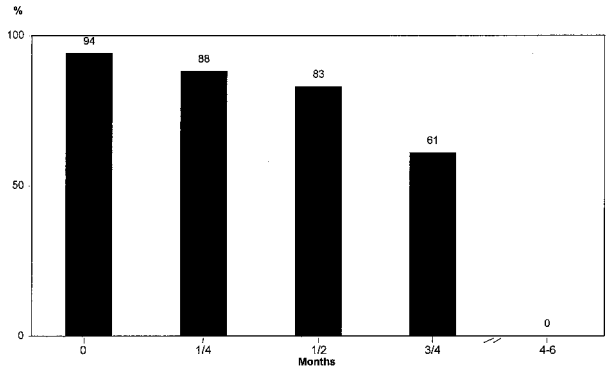

Fig. 7. Proportion of patients who achieve complete recovery as a function of time of beginning recovery after the onset of paresis.

third week had a significantly worse outcome ($p < 0.001$). It is clear that the time of beginning remission is highly significant to the prognosis.

Age of patients. Age is another parameter that influences the final result (Fig. 8). Children aged ≤ 14 years had the most favorable prognosis, with 90% achieving full recovery. Patients aged 15–29 years had a fairly good chance of recovery (84%). The chance of a full recovery was reduced for patients aged between 30 and 44 years (75%). Above the age of 45 years, the chances of recovery diminished significantly (64%). Above the age of 60 years, only about one-third of patients will experience the return of normal function. The influence of age on the final outcome is therefore highly significant ($p < 0.001$).

Postauricular pains. As noted above, postauricular pains were registered in 52% of all cases of Bell's palsy. A total of 78% of patients with no pain regained normal function, as opposed to only 64% of patients with pain ($p < 0.001$).

The prognostic value of topographical tests. It must be stressed that the examination of taste, stapedius reflex and tear flow or nasolacrimal reflex (Table VIII) should be performed very carefully and always in exactly the same way or else the comparison of results is meaningless. The results should not depend on the person performing the tests.

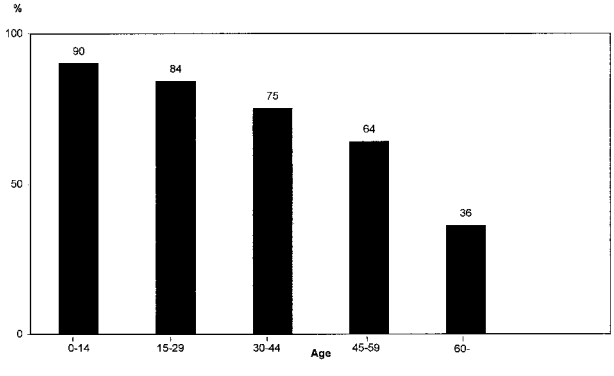

Fig. 8. Age distribution of patients who achieve complete recovery.

Taste. The taste test according to Boernstein (36) is semiquantitative and based on recognition of four basic tastes—sweet, salt, sour and bitter—at three different concentrations. Initial taste examination showed that 83% of patients had partially reduced or abolished taste while 12% had normal taste. Final taste examination showed that 80% of patients had regained normal taste function. Taste function and the muscular function of the face normalized at approximately the same time.

Stapedius reflex. The stapedius reflex is an acoustic facial reflex provoked on both sides by one-sided sound stimulation (37). Initially, 72% of patients had a reduced or abolished reflex and only 22% had a normal reflex. When remission occurs, stapedius reflex will usually return 1–2 weeks before visual function of the facial muscles can be confirmed. Normal function was restored in 86% of patients.

Tearing or nasolacrimal reflex. The nasolacrimal reflex passes from the nasal mucosa to the superior salivary nucleus and thence to the secretory fibers, with the facial nerve ending in the lacrimal gland. In this study a modification of Schirmer's test II (38) was used, which is based on measurement of tear flow for 1 min. Stimulation with benzene is carried out for 30 s and measurement is performed by using filter paper placed in the lower fornix. At the end of the test the length of the soaked strip of filter paper is measured on both sides. The difference between the 2 sides is $< 20\%$ in 95% of normal persons. Initially, 11% of patients had partially reduced or abolished tearing. The final result showed that 97% of patients achieved normal tearing. In comparison with taste and stapedius reflex testing, tearing became normal in a surprisingly high proportion of patients.

Figure 9 shows the prognostic value of the three topographical tests. The patients were divided into two groups: one group with normal facial muscle function and the other with muscular sequelae. Comparison of the results of the initial taste tests for the groups with and without sequelae showed that 91% and 80% of patients, respectively had partially reduced or abolished taste ($p < 0.001$). Concerning the stapedius reflex, it was found that 91% and 63% of patients, respectively had a reduced or abolished reflex ($p < 0.001$). The nasolacrimal reflex is also a reliable prognostic indicator because 27% and 5% of patients, respectively had abolished or reduced lacrimal function ($p < 0.001$). In conclusion, all three of the topographical tests provide reliable prognostic information.

Table VIII. *Results of the initial and final examinations of taste, stapedius reflex and nasolacrimal reflex*

Test	Reduced or abolished function		None assessible		Normal function	
	n	%	n	%	n	%
Taste						
Initial	1,419	83	111	7	171	10
Final	236	14	111	7	1,354	80
Stapedius reflex						
Initial	1,221	72	108	6	372	22
Final	122	7	122	7	1,471	86
Nasolacrimal reflex						
Initial	192	11	27	2	1,482	87
Final	29	2	27	1	1,645	97

Sequelae. As mentioned before, 71% of patients regain normal function of their facial muscles after an idiopathic paresis. The remaining 29% of patients suffer from varying degrees of sequelae. It is extremely difficult to describe the sequelae accurately and to group the degree of sequelae; however, it should be borne in mind that the daily discomfort of sequelae is a significant problem for the patients. Today, there are at least 10 systems available for facial nerve grading but none of these systems is ideal (39, 40).

The existing systems include gross scales, regional systems and specific scales. One of the biggest problems with grading systems is finding a balance between an exact description of the sequelae and minimizing the number of groups into which the patients are classified. The scale should combine the main parameters (paresis, contracture and associated movements) based on specific definitions. An ideal system is one with high specificity and an acceptable sensitivity. Before proposing my gross scale the first 200 of the patients in this study were examined 3 times. The scale emerged from an examination of these 200 patients who suffered from peripheral facial nerve palsy with different grades of reduced function and other sequelae. It should be stressed that the scale includes all types of motoric dysfunction. Other secondary defects, such as crocodile tears, decreased tearing, taste disturbances and stapedius muscle problems, are registered and should be added to the other sequelae to find the total number of sequelae. My scale is a modification of that of Botman and Jongkees (41).

The factors determining the degree of sequelae are paresis, contracture and associated movements (also known as mass movements or synkinesis). The definition of synkinesis is an involuntary movement accompanying a voluntary one. The final cosmetic result depends on a combination of these three elements.

Table IX shows the grade of palsy. Definitions of grades 0–IV are given in Table X. Contracture causes a narrowed palpebral fissure on the involved side when the face is in repose. The corner of the mouth occurs higher on the affected side and the folds of the face are abnormally deep; in particular the nasal labial fold is very marked. Associated movements are found in the muscles of the eye, cheek and mouth and more seldom in the forehead. Associated movements are visible when the patient tries to close his/her eye and smiles involuntarily or vice versa. Associated movements often cause more cosmetic inconvenience to a patient than a slight paresis or contracture. The three parameters should be combined to give an accurate description of sequelae.

The degree of recovery for all patients is demonstrated in Fig. 10, which shows that 71% recovered completely, 12% had slight sequelae, 13% had moderate sequelae and only 4% had severe sequelae. It should be stressed that no patients became paralyzed. In summary, 83% of patients had a good recovery and 17% a bad recovery without any kind of treatment.

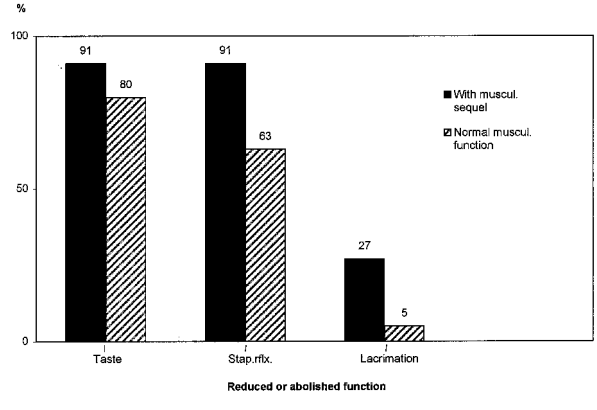

Fig. 9. Proportions of patients with and without muscular sequelae who had reduced or abolished function in the three topographical tests.

Table IX. *Grades of palsy in the Peitersen grading system*

Grade	Degree of palsy	Description of palsy
0	None	Normal function
I	Slight	Only visible when patient grimaces
II	Moderate	Visible with small facial movements
III	Severe	Function just visible
IV	Complete	No function

Figure 11 shows the types of initial damage of the facial nerve. A total of 85% of patients experienced different types of damage, such as neuropraxia, axonotmesis, neurotmesis and partial degeneration. For all these patients recovery began within 3 weeks after the onset of palsy. Only 15% of patients suffered total degeneration of the nerve, with recovery beginning ≥ 3 months after the onset of palsy.

Figure 12 demonstrates the final outcomes after recovery. As mentioned above, 83% of patients had a good outcome and 17% a bad outcome.

Comparison of the results shown in Figs 11 and 12 shows that there is a very clear connection between early pathology in the facial nerve and the final outcome. A total of 85% of patients had a "mild pathology" and 83% achieved a fair final result. A total of 15% of patients had a "severe pathology", namely total degeneration of the nerve. All of these patients (and 17% in total) were in the group with severe sequelae.

A comparison of my scale of sequelae and that designed by House and Brackmann (40) is possible to some extent. If their grades IV and V are combined then the new group is almost identical to my grade III (Table XI). The number of Bell's palsy patients in this group is very small and to the best of my knowledge it is very difficult to distinguish between House and Brackmann's grades IV and V.

The distribution of sequelae is listed in Table XII. Paresis is the commonest sequela but not the most uncomfortable for the patients. Associated movements and contracture were found in 16% and 17% of the sample, respectively. Both sequelae cause more inconvenience for the patient than a slight paresis. The most troublesome sequelae for patients are the associated movements. Contracture gives the patient a feeling of stiffness in the muscles of the face. In five patients with disfiguring contractures biopsies were taken from the musculus orbicularis oris on both the paretic and normal sides. Microscopy showed a reduction in the number and size of the muscle cells and an increased amount of connective tissue and fat on the paretic side. As can be seen from Table XII, crocodile tears and dry eyes are very rare, with both

Table X. *Description of sequelae associated with grades 0–IV of palsy in the Peitersen grading system*

Grade	Palsy	Contracture	Associated movements
0	None	None	None
I	Slight	Just visible (<1 mm)	None
II	Moderate	Clearly visible	Visible
III	Severe	Disfiguring	Marked
IV	Complete	None	None

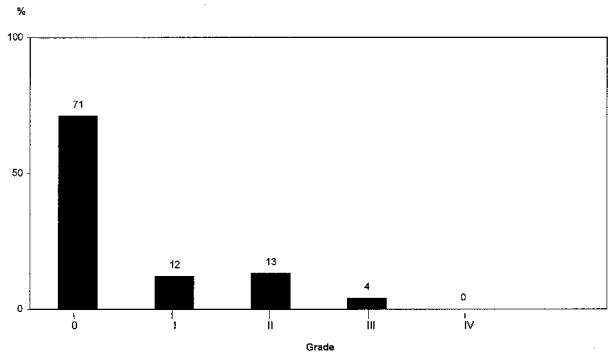

Fig. 10. Distribution of degree of recovery for the patients with Bell's palsy.

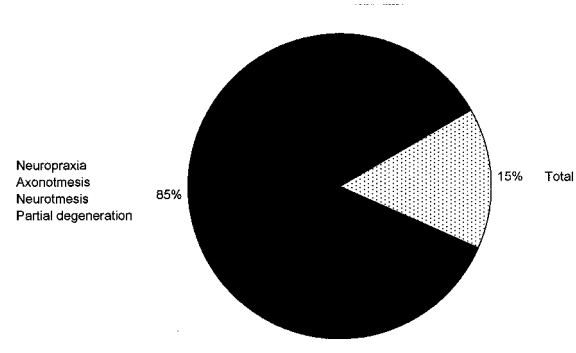

Fig. 11. Distribution of types of initial damage of the facial nerve.

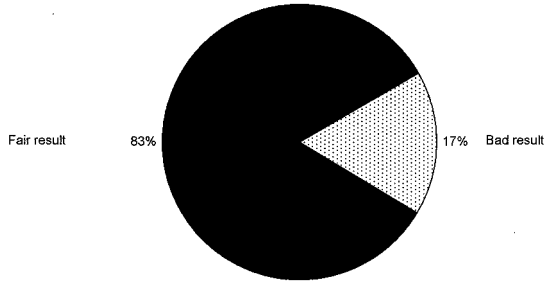

Fig. 12. Distribution of final outcomes after recovery.

Table XI. *Comparison of the Peitersen and House and Brackmann grading systems*

Peitersen		House and Brackmann		Patients with Bell's palsy	
Grade	Degree of palsy	Grade	Degree of palsy	n	%
0	None	I	None	1,202	71
I	Slight	II	Mild dysfunction	211	12
II	Moderate	III	Moderate	220	13
III	Severe	IV &V	Moderate/severe and severe	68	4
IV	Complete	VI	Total paralysis	0	0

Table XII. *Distribution of sequelae of Bell's palsy in the total patient sample*

Sequela	%
Paresis	29
Associated movements	16
Contracture	17
Crocodile tears	2
Dry eye	2

sequelae occurring in only 2% of all Bell's palsy patients.

The prognosis for Bell's palsy

The prognosis depends to a great extent on the time at which recovery begins. Early recovery gives a good prognosis and late recovery a bad prognosis. Age is another parameter that influences the final result. Young people have a good prognosis and old people a worse prognosis. Normal taste, stapedius reflex and tearing give a significantly better prognosis than if these functions are impaired. The prognosis for Bell's palsy is significantly negatively affected if postauricular pains occur. Finally, the recovery profile is significantly better for patients with incomplete paresis compared to those with complete paralysis.

PERIPHERAL FACIAL NERVE PALSIES CAUSED BY HERPES ZOSTER

The Copenhagen Facial Nerve Study includes 116 cases of untreated peripheral facial nerve palsy caused by herpes zoster. The combination of this viral infection with facial nerve paralysis is not unusual: in this sample the ratio of idiopathic facial nerve palsy to herpes zoster palsy was 15:1.

Herpes zoster oticus generally has a poor prognosis and many patients are left with permanent facial nerve sequelae. The syndrome was described by Miehlke in 1904 (14), but is better known as Ramsay Hunt syndrome. Hunt (42) described the pathological findings in the geniculate ganglion in 1907. However, the infection is not only localized to the ganglion but is, according to Miehlke (43), a generalized muco-dermato-polyneuro-encephalo-myelo-meningitis disease.

In this study the 116 patients with peripheral facial nerve palsy caused by herpes zoster comprised 51 males and 65 females. The age of the patients ranged from 11 to 89 years. Fig. 13 demonstrates the age distribution of these patients in comparison with that of the underlying population. There is a maximum incidence of peripheral facial nerve palsy caused by herpes zoster above the age of 45 years; only a few children and young people suffer from it.

Table XIII shows that 102 (88%) of the herpes zoster patients had paralysis and only 14 (12%) suffered from incomplete paresis. Compared with Bell's palsy patients, herpes zoster patients have more severe lesions. Table XIII also shows that there was a high incidence of associated symptoms such as hearing loss (73%) and vestibular disturbances (64%). In 55% of the patients combined cochleovestibular lesions were found. The very typical hearing loss is a sensory neural high-tone loss and as a rule is non-reversible. More severe hearing losses can be seen but anacusis has not been observed.

The results of topographical testing are shown in Table XIV. It is evident that the number of sequelae is much higher in the herpes zoster patients than in the Bell's palsy patients.

The diagnosis of herpes zoster oticus is to a large extent based on clinical observations. Table XV shows the localization of the herpes zoster vesicles in all 116 patients. Only two-thirds of the patients had blisters localized in the ear, proving that it is necessary to inspect the head, neck, oral cavity, pharynx and thorax. Another problem concerns when the vesicles appear. There are no diagnostic problems if the vesicles occur before the facial nerve palsy. However, if the vesicles do not occur first then the paresis could be diagnosed as Bell's palsy. In this sample, paresis occurred after the vesicles in 60% of cases, in 25% they occurred simultaneously and in 15% the vesicles occurred after the facial nerve palsy.

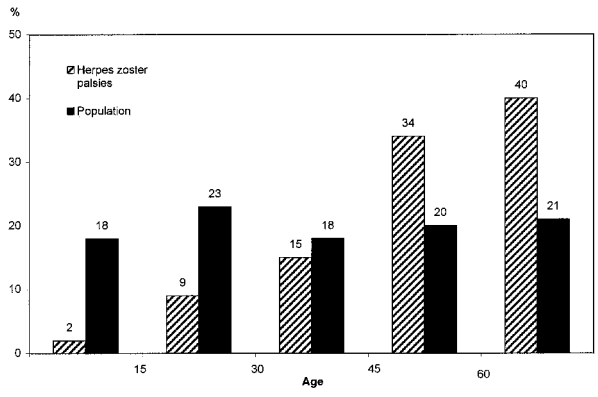

Fig. 13. Age distribution of patients with herpes zoster in comparison with that of the underlying population.

The diagnosis of herpes zoster was based on clinical observations and determination of specific antibodies in serum and CSF. A lumbar puncture was performed in 20 consecutive patients. CSF showed a raised protein level of 0.8–1.8 g/l and pleocytosis of $40–300 \times 10^6$ leukocytes/l. (Normal protein levels are defined as < 0.5 g/l for young and middle-aged patients and < 0.7 g/l for elderly patients. Pleocytosis is defined as $> 3 \times 10^6$ leukocytes/l.) Positive antibodies were also found in all patients. Re-examination was performed in six patients 3–6 months after the initial examination. All patients still had raised protein levels and pleocytosis but the degree of abnormality had decreased.

The prognosis for restoration of facial nerve function in herpes zoster patients is poor (Fig. 14). Only 21% achieve normal function, 25% have mild sequelae only, 26% have moderate sequelae, 24% severe sequelae and 4% no function at all. The recovery profile is fair for 46% of patients and bad for 54%.

When this study was finished, treatment of herpes zoster palsies was started. Only 28 patients were included in this program. A 6-day treatment with acyclovir, 10 mg/kg i.v. every 8 h, was used (44). The results were not encouraging. The recovery for the treated group did not differ significantly from that for the untreated group. No effect could be demonstrated on hearing loss, but it was obvious that tinnitus and vertigo decreased within a few days. However, the patient sample was too small for definitive conclusions to be drawn.

Table XIII. *Distribution of herpes zoster palsies*

Paresis	n	%
Complete	102	88
Incomplete	14	12
Hearing loss	85	73
Vestibular lesions	74	64
Combined cochleovestibular disturbances	64	55

Table XIV. *Final results of topographical tests for Bell's palsy patients and herpes zoster patients. Values shown represent percentages of patients with normal function in the tests*

Test	Patients with herpes zoster	Bell's palsy patients
Taste	43	79
Stapedius reflex	46	87
Nasolacrimal reflex	87	96

Table XV. *Localization of herpes zoster vesicles in the 116 patients with herpes zoster*

Nerve/skin innervation	%
Concha and external canal	66
Trigeminal	15
Glossopharyngeal	5
Vagus	4
Cervical nerves 2 and 3	7
Thorax	3
Total	100

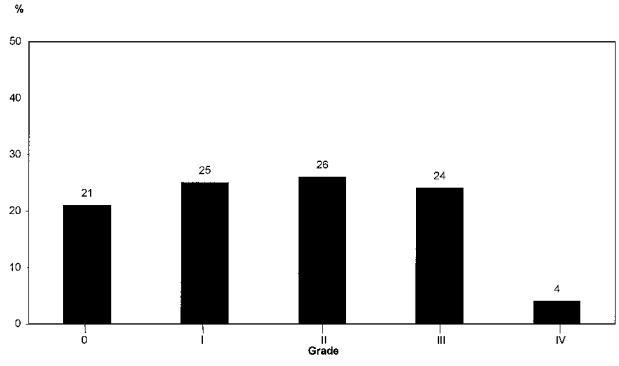

Fig. 14. Distribution of degree of recovery for herpes zoster patients.

PERIPHERAL FACIAL NERVE PALSIES AND DIABETES MELLITUS

Diabetes mellitus is a generalized metabolic disorder characterized by elevation of the blood glucose level. The disease causes damage to the vascular system and this insufficiency produces very common central and peripheral nervous system disorders. The nerves innervating the eye muscles are the most frequently affected, followed by the facial nerve. The paresis is unilateral and recurrent paresis is common; however, bilateral peripheral facial nerve palsies have also been

described. This study included 76 diabetics (42% male; 58% female) with peripheral facial nerve paresis. The patients were aged between 21 and 82 years and all were treated with insulin. The ratio of idiopathic palsy to palsy in diabetics was 22:1.

Of the 76 patients, 47 (62%) suffered from incomplete paresis and 29 (38%) from complete paralysis. It was a surprise that the majority of these patients (almost two-thirds of them) had incomplete paresis and an even greater surprise that the recovery for these patients was very poor, with only 25% achieving normal facial muscle function. The explanation for the poorer degree of recovery of facial nerve function in these patients is undoubtedly the underlying disease of diabetic polyneuropathia. In Denmark diabetes is estimated to affect 3–4% of the population.

PERIPHERAL FACIAL NERVE PALSIES IN CHILDREN

Of a total of 2570 patients with peripheral facial palsy, 349 were aged < 15 years. The etiology of these patients is shown in Table XVI; for a review see May (21). It can be seen that Bell's palsy, which includes idiopathic palsy, comprises about one-third of the cases and that the largest group of patients comprises neonates. The group of multiple malformations includes the real congenital palsies (neonatal age n = 169). The subject of congenital versus birth traumatic palsies will be discussed later. There are very few patients in the other groups, with the exception of bilateral palsies, which are probably caused by viral infections. Eight of these cases occurred during the same month. A slight fever, headache and, in five of the cases, vomiting were reported before the facial paresis occurred. Three of the patients showed bilateral paresis within 24 h. Lumbar puncture was performed in five cases but CSF was normal and Echo and Coxsackie virus investigations were negative. In the other three cases there were intervals of 3–8 days between the occurrence of paresis on both sides. All the children regained normal facial function.

Table XVI. *Etiology of peripheral facial nerve palsies in children (<15 years)*

Etiology	n
Bell's palsy	138
Neonatal age	169
Bilateral palsy	16
Acute otitis	7
Temporal bone fracture	6
Herpes zoster	2
Chronic otitis	2
Infectious mononucleosis	2
Leukemia	2
Cholesteatoma of the middle ear	1
Melkersson–Rosenthal syndrome	1
Malignant lymphoma	1
Pemphigus	1
Smallpox vaccine sequelae	1
Total	349

Table XVII illustrates that neonates with presumed birth trauma are the largest group. In 33 patients all branches were affected and in 68 only the marginal mandibular branch of the facial nerve was affected. The marginal mandibular branch is the most vulnerable of all branches. This branch innervates the depressor muscle group of the lower lip and in cases of paresis this will result in a straight lip on the paretic side.

This particular group of patients is continually under discussion with regard to whether paresis is congenital, due presumably to aplasia of the facial nucleus, or is perhaps due to birth trauma. For several reasons many supporters of the first theory have now abandoned it. The primary reason being that, if the theory of congenital paresis holds, then function would not be likely to improve. Improvement has, however, been seen in a certain number of cases. Second, from surgical experience and from knowledge of Bell's palsy it is known that the mar-

Table XVII. *Localization of facial nerve paresis in neonates with presumed birth injuries and distribution of degree of recovery as a function of localization of paresis*

n	Branches	Paresis	000	Recovery		
			n	No	Yes	Normal
33	All	Complete	7	5	2	0
		Incomplete	26	1	25	9
44	>1	Complete	12	9	3	1
		Incomplete	32	1	31	14
68	M.m.br.	Complete	41	39	2	0
		Incomplete	27	4	23	11
		Total	145	59	86	35

M.m.br. = marginal mandibular branch.

Table XVIII. *Congenital abnormalities and facial nerve palsies in neonates*

Abnormality	n
Treacher Collins syndrome	16
Moebius syndrome	2
13-trisomy (Patau's syndrome)	1
18-trisomy (Edward's syndrome)	1
Multiple defects	4
Total	24

ginal mandibular branch is the most vulnerable and that its regeneration is the poorest in Bell's palsy patients, with $\approx 10\%$ never recovering function of this branch. Third, the number of birth traumatic palsies has decreased to $\approx 15\%$ during the last 25 years as a result of improved obstetric techniques. Finally, electrical tests, such as electromyography and EnoG, allow one to distinguish between peripheral and central lesions. However, babies untouched by hands and forceps in utero can still have paralysis of the marginal mandibular branch. Experience shows that if the marginal mandibular branch is paralyzed then function will never be restored. Details are shown in Table XVII. Table XVIII shows some congenital abnormalities and facial nerve palsies in neonates. The majority of cases suffer from Treacher Collins syndrome. The other children had multiple defects or chromosomal abnormalities with one-sided or bilateral palsies.

Figure 3 demonstrates the age distribution of patients with Bell's palsy in comparison with that of the underlying population. The disease is significantly less common below the age of 15 years ($p < 0.001$). Furthermore, children have the most favorable prognosis, with 90% achieving full recovery (Fig. 8).

PERIPHERAL FACIAL NERVE PALSIES IN PREGNANCY

Peripheral facial nerve palsy is uncommon in pregnant women. The ratio of idiopathic facial nerve palsy in women to palsy in pregnancy was 19:1 in this study. A comparison of recovery between non-pregnant females aged 15–44 years and pregnant women showed that normal function was obtained in 80% and 61%, respectively. The prognosis of peripheral facial nerve palsy for pregnant women is significantly worse than that for non-pregnant women of the same age ($p < 0.001$).

PERIPHERAL FACIAL NERVE PALSIES OF DIFFERENT ETIOLOGIES

Figure 15 shows a comparison of the final results of facial nerve palsies of different etiologies. It can be seen that 71% of the idiopathic group, 61% of pregnant women, 21% of herpes zoster patients, 25% of diabetes mellitus patients and 24% of polyneuritis patients recovered completely. The majority of patients suffering from B. burghdorferi infection were included in the polyneuritis group. These figures lead to the conclusion that idiopathic palsy has the best prognosis of all the types of peripheral facial nerve paralysis.

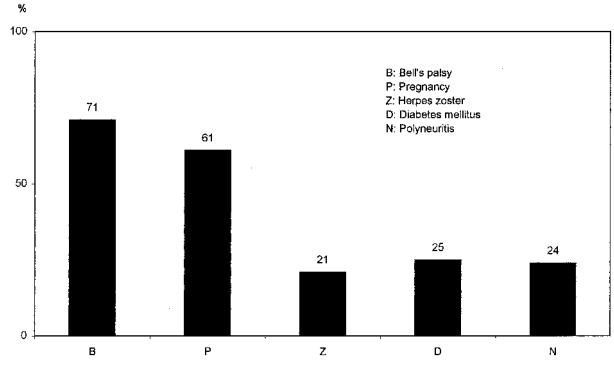

Fig. 15. Proportions of patients who achieved complete recovery from facial nerve palsies of different etiologies.

AN EVALUATION OF TREATMENT OF BELL'S PALSY

Can Bell's palsy be treated?

As mentioned in the Introduction, Friedreich was the first doctor to attempt to treat patients suffering from idiopathic peripheral facial nerve palsies. During the last two centuries an unknown, but large, number of these patients have been treated with almost every kind of medicine, physiotherapy, electrical stimulation and surgery. More than 1,000 papers have been published and the conclusions, with very few exceptions, have been that the patients benefited from the treatment and that the authors were convinced of its efficacy, even though there was no real proof.

Fortunately, reviews of the literature on the treatment of facial paralysis exist. Cleveland gave a review covering the period 1932–38 (45) and Ghiora and Winter (45) reviewed the literature on conservative treatment of Bell's palsy between 1939 and 1960. These reviews make very interesting reading and it is stressed that the evaluation of therapy is made difficult by the usually high percentage of spontaneous and complete recovery. Many patients show signs of returning function as early as 10 days after onset, even without treatment. Conservative treatment is of course designed to reduce edema, ischemia, congestion and compression, and thus to prevent total degeneration. In the following sections a short overview of treatment methods will be presented.

Thermal methods

In the older literature the majority of authors advocate conductive, radiative and convective heat transfer in order to achieve vasodilatation. Vasodilatation may be both local and reflex in nature and it is logical to attempt vasodilatation in view of the acceptance of vasoconstriction as the major pathogenic factor. Furthermore, experience has shown the soothing effect of heat treatment in patients with Bell's palsy. However, some authors assert that heat treatment may increase edema and thus is inadvisable. Instead of heat treatment, some patients were treated by applying ice over the mastoid region with the aim of relieving edema. However, according to the ischemia theory, such treatment would increase vasoconstriction and work against the intention of vasodilatation (45). No controlled clinical trials have been published in this area and heat therapy may be considered to be a form of psychotherapy.

Electrotherapy

Electrotherapy is one of the most controversial subjects in the treatment of peripheral facial nerve paralysis. Some authors have advocated electrotherapy but others have criticized it. According to Ghiora and Winter it is useless and possibly even dangerous, because it may cause contractures (45). Mosforth and Taverner (46) reported a controlled trial of the value of galvanic stimulation in the management of 86 Bell's palsy patients. The authors concluded that although no significant advantage could be demonstrated by the use of galvanic stimulation the presence of contracture was not related to the mild electrical treatment. However, Williams (47) cautioned against exaggerated forms of physiotherapy, especially electrical stimulation, as this kind of therapy may lead to permanent contractures. As electromyography has demonstrated that even a denervated muscle will preserve its function for at least a year, it is difficult to see what physiotherapy will contribute to recovery. If reinnervation takes place it will occur within a year and the innervated muscles will recover their function regardless of physiotherapy. On the other hand, if denervation is permanent, no amount of physiotherapy will prevent muscular degeneration.

Different electrical stimulation apparatuses, some of them very sophisticated, were constructed during the last century. Galvanic and faradic stimulation were used.

This study included 28 patients with recurrent paresis on the opposite side. The first paresis in these patients had been treated with intensive electrical stimulation. The second paresis was not treated but was followed until normal function was obtained or for up to 1 year. A comparison of the 2 sides of the face showed that 23 patients had more marked contractures on the treated side. The grade of sequelae was II–III on the treated side and I–II on the untreated side but, as stressed by Langworth and Taverner (32), the development of contractures requires degeneration.

Massage

The value of massage is to produce hyperemia and maintain tonus of the facial muscles. Different opinions exist concerning the efficacy of massage, but again no significant studies have been published. Massage may be considered to be a form of psychotherapy (45).

Facial exercise

For many years facial exercises have been recommended for peripheral facial nerve palsy patients with both complete and incomplete paresis. The patient should stand in front of a mirror and watch the face while raising the eyebrows, gently closing the eyes, wrinkling the nose, whistling, blowing out the cheeks and grinning. These facial exercises should be performed twice a day (21, 45, 48). Although the effect of facial exercises has not been statistically evaluated, patients appreciate the exercises to a very great extent. However, they should be considered a form of psychotherapy according to Wolferman (49).

Cervical sympathetic block

Blocking of sympathetic pathways may relieve vasodilatation of the vasa nervorum to the facial nerve. This may be applied to the stellate ganglion using procaine. Korkis (50) claimed satisfactory results, but no controlled trials have been reported. Fearnley et al. (51) found no significant benefit from the use of this method.

Surgery

In 1932 Balance and Duel (12) advocated a transmastoid decompression operation in patients with Bell's palsy. In the years that followed the number of operations increased dramatically, but the well-known decompression surgeons Cawthorne (13), Jongkees (15), Miehlke (14) and Kettel (17) did not try to explain their treatment results. According to Jongkees (52), Kettel (53) and Miehlke (14) the indication for surgery was paralysis lasting 2 months. In their experience patients achieved function $\approx$ 1 month after the operation. However, this study has documented that the operation is useless and that spontaneous regeneration results in regained function. As mentioned before, indication for surgery was based on a mechanical way of thinking and furthermore there is not sufficient material to prove the efficacy of decompression surgery for Bell's palsy.

Fisch (16, 54) recommended total decompression of the facial nerve from the styloid foramen to the internal ear canal if the electroneurographic degeneration exceeded 90% within 6 days after the onset of palsy. This indication is based on his so-called ''bottleneck theory''. The present patient sample is too small to fulfill the requirements for randomization (55, 56).

Yanagihara et al. (57), who believe in transmastoid decompression, have sought to prove the efficacy of the operation. Their patient sample included 101 Bell's palsy patients initially treated with steroid but with denervation exceeding 95% and, at early examination, a function equivalent to House and Brackmann's grade V or VI. Surgery was performed during three different periods from the onset of palsy: (i) within 1 month; (ii) during the second month; or (iii) after 2–4 months. The best results were obtained in the first group, i.e. those with the earliest surgery. Unfortunately, information about the time from the onset of palsy to the first sign of recovery is not available. This study shows that patients with no function after 3 weeks undergo total degeneration and begin recovery after 3–5 months. The time of operation and the time of spontaneous recovery coincide. Information about the period between the operation and the beginning of recovery would have been valuable but is also not available. There was an age difference between the surgery and control groups and furthermore the number of patients was too small to permit significant statistical evaluation. It is impossible to explain how decompression can help a patient 2–4 months after the onset of palsy, when the facial nerve is undergoing regeneration. The trial of Yanagihara et al. does not provide evidence of the efficacy of the decompression operation in Bell's palsy patients who were initially treated unsuccessfully with steroids.

Drug therapy

Despite the appearance of many publications on drug therapy during the period 1930–60, there were few adequate therapeutic trials. Initially, vasodilators were mainly used, based on the etiologic factor of ischemia of the vasa nervorum of the facial nerve. Later on steroids were introduced in an attempt to influence a possible non-specific acute inflammatory reaction.

Vasodilators

Different authors have used a variety of vasodilator drugs. Despite pharmacological observations in normal persons and animals, the actual value of many drugs in peripheral vascular disorders is uncertain. The drugs used were histamine, procaine, nicotinic acid, nitrites and papaverine (45). Alarming side-effects associated with histamine and i.v. procaine were not unusual and flushing resulted from the administration of large doses of nicotinic acid. Korkis (50) concluded that it was possible that the vasodilated vessels leading to the affected nerve caused further swelling of the nerve within the bony canal, thereby aggravating compression and secondary ischemia. In view of the danger and doubtful benefit of many of these drugs in many peripheral vascular disorders, there seems little basis for recommending vasodilators.

Prednisone

Taverner (58) in 1954 was the first to design a controlled treatment trial of steroids but unfortunately the number of patients was too small to permit a significant statistical evaluation. Attempts to treat Bell's palsy with steroids changed in the 1970s. The publication of Adour et al. (59) in 1972 concerning prednisone treatment for idiopathic facial paralysis was a milestone in the treatment of Bell's palsy but unfortunately the double-blind protocol was abandoned in 1970, because the placebo-treated patients demanded steroids. What these patients were really seeking was the euphoric side-effect of prednisone. The trial included 194 treated and 110 untreated Bell's palsy patients. The controls were retrospective and the study was not blind or randomized. The NET was used to demonstrate the absence of complete nerve denervation in all treated patients but, as stressed by Blumenthal (29), Groves and Gibson (30), Laumans (31) and Wolferman (49), this test is not reliable. The description of sequelae was not exact, because patients with only 76% function of the facial muscles obtained a maximum points score of 10, so that it is impossible to find out how many patients regained normal function. However, the conclusion was that the treated group experienced fuller recovery and less severe complications.

After the initial publication of Adour et al. (59), several series of treatments with prednisone for Bell's palsy were designed, but almost all of them were of unsatisfactory quality. Nevertheless, the majority of authors claimed that they had shown steroids to be beneficial to a statistically significant degree. One exception was the report of May et al. (60) in 1976. They concluded that there was no proven efficacious treatment for Bell's palsy. Unfortunately the material was too small to permit a valid statistical analysis. Burgess et al. (61) described the problems in 1984, but more recently designed trials do not fulfill the requirements for a clinical study (Table XIX). In a comprehensive review of the literature in 1987, Stankiewicz (62) concluded that ''A definitive statisti-

cally valid study considering the benefit of steroids in the treatment of idiopathic facial nerve palsy has yet to be performed''. Prescott (63) in 1988 could not demonstrate any effect of prednisone treatment in 879 patients.

Based on the conclusion of Stankiewicz, Austin et al. (64) in 1993 concluded that a randomized controlled study to evaluate the efficacy of oral steroids in the treatment of idiopathic facial nerve palsy was necessary. They published a well-designed study comparing the use of oral prednisone versus placebo in the treatment of idiopathic facial nerve palsies. It was obvious that only one objection could be raised against the trial, albeit a very serious one. There were too few patients in the study ($n = 76$) to enable statistically significant conclusions to be drawn (61). Nevertheless the conclusion was that patients treated with prednisone had less denervation and a significant improvement in the facial grade of recovery than the placebo-treated patients. No differences were found between the two groups in terms of the time period for recovery, the percentage of patients with synkinesis and the percentage of patients with crocodile tears. Both synkinesis and crocodile tears are caused by degeneration and so it is not easy to see how the treatment could help when the time of recovery and the percentage of patients with synkinesis were the same in the prednisone- and placebo-treated groups.

Shafshak et al. (65) performed a prospective study on 160 unilateral non-recurrent Bell's palsy patients treated with prednisone at suggested sufficient doses. The study was randomized, but there was a male predominance (129 males, 31 females) and placebo was not used. The number of patients was less than half that required to draw statistically significant conclusions (61). Statistical comparison between the control group and the steroid subgroups revealed that patients who started prednisone intake within 24 h of onset had a significantly better recovery than those in the control group, but only 23 patients benefited in this way. Furthermore, there was no significant difference in outcome between patients in the control group and those who started prednisone intake > 24 h after onset. The study showed that only 23/93 patients treated benefited from the medication.

Table XIX. *Requirements for documentation of therapy for peripheral facial nerve palsies*

Prospective study
Sufficient number of patients
No selection of patients
Clear inclusion and exclusion criteria
Randomized study
Double-blind placebo study
Follow-up until restoration of normal function or for 1 year
Exact description of sequelae
Adequate statistical analysis
Conclusions based on the results

Ramsey et al. (66) in 2000 conducted a meta-analysis to evaluate facial recovery in patients with complete idiopathic facial nerve paralysis by comparing outcomes of those treated with prednisone therapy with outcomes of those treated with placebo or with no treatment. A total of 47 trials were identified; 20 of these trials were excluded because they were retrospective and 24 prospective studies were excluded for a variety of reasons such as lack of outcome, multiple medical or surgical treatments or a steroid dose that did not meet the inclusion criteria. Therefore only three trials were assessed to meet the inclusion criteria, namely those by May et al. (60), Austin et al. (64) and Shafshak et al. (65). The meta-analysis of Ramsey et al. (66) is a considerable piece of work but seems to contain some illogical statements. Although the statistical analysis is extremely thorough, the data were manipulated: initially three trials (60, 64, 65) were included but later the trial of May et al. (60) was excluded because of the small sample size that showed a worse outcome with the use of steroid treatment. Consequently, a pooled analysis from the other two trials was presented. A minor problem with the meta-analysis is the incorrect number of subjects mentioned by May et al. (60) and by Austin et al. (64). A more serious problem is that the numbers of patients in all three trials are too small and even a combination of the three samples does not give a sufficient number of patients, according to Burgess et al. (61).

The meta-analysis of Ramsey et al. (66) concludes that ''Corticosteroid treatment provides a clinically and statistically significant improvement in recovery of function in complete idiopathic facial nerve paralysis''. This conclusion is based on what is called ''marginal significance'' in the trial of Austin et al. (64) and ''significance'' in the study of Shafshak et al. (65). However, Shafshak et al. showed only a better recovery for the subgroup of 23 patients who began steroid intake within 24 h after the onset of palsy. Therefore only 23/93 patients treated benefited from the medication. To the best of my knowledge I cannot see that the conclusion of the meta-analysis is correct because there is a marked discrepancy between the data analyzed and the conclusion in the abstract.

Antiviral drugs

In 1996 Adour et al. (67) reported that combined treatment with prednisone + acyclovir restored 92% of patients to normal function but, as in these au-

thors' prednisone study from 1972 (59), a special grading system was used. Furthermore, 13% of patients developed contracture and synkinesis, so that the proportion of patients who achieved a fair result should be reduced to 87%; again it is impossible to see how many patients obtained full recovery. Furthermore, the patients in the study of Adour et al. were highly selected, with 80% having incomplete paresis. It should be stressed that $\approx 95\%$ of initially incomplete paresis patients obtain normal function without treatment. The follow-up used in this study (4 months) was too short. After 4 months it is not possible to assess sequelae because contracture and synkinesis have not developed to their full extent. As mentioned above it is necessary to follow these patients for 12 months until a steady state is attained. The number of patients in the study was also too small to permit statistical significance (61). Adour et al. (67) found contracture and synkinesis in 13% of the acyclovir + prednisone-treated group and in 28% of the placebo (prednisone-treated) group. In 1972 Adour et al. (59) had found contracture and synkinesis in 15% of the prednisone-treated group and 21% of the untreated group. It is difficult to explain the difference between the prednisone-treated groups in 1972 and 1996. The placebo (prednisone-treated) group in the 1996 study had a worse outcome (28%) than the untreated group in 1972 (21%) and this discrepancy is inexplicable. Despite this Adour et al. emphasized that the NET did not show degeneration in the steroid-treated group; they described contracture and synkinesis in 15% of patients and as a matter of fact development of contracture as well as synkinesis requires severe degeneration (68).

A comparison between the present study, in which patients were not treated, and that of Adour et al. (67) shows no difference in the proportion of patients achieving a fair outcome: 83% and 87%, respectively. As stressed above, the proportion of patients with incomplete paresis in the 2 studies was 30% and 80%, respectively.

Combined therapy

Some authors regard Bell's palsy as an emergency that should be treated using all available measures against possible multiple factors. Therefore a combination of medicine, electrical stimulation, physical therapy and sometimes even decompression operation has been used. Sittel et al. (69) published a retrospective trial in 2000 that included 334 patients suffering from sudden facial paralysis of unknown cause. Only 239 patients were recorded as having been treated with a drug cocktail consisted of prednisone, dextran and pentoxifylline. The term antiphlogistic–rheologic infusion therapy (ARIT) has been coined to describe this regimen. The majority of patients 173 (72%) had incomplete palsy and only 66 (28%) had complete paralysis. Patients with incomplete palsy obtained full recovery in 98% of cases, a result which does not differ from the result (94%) obtained without treatment in this study. Patients with complete paralysis regained normal function in 77% of cases, as opposed to 61% of patients in this study. It is surprising that in the study of Sittel et al. (69) diabetics obtained the same final outcome as normal persons. In the present study it was found that only 25% of diabetics regained normal function and, furthermore, they had recurrences more often.

When comparing trials of treatment of facial palsies it is generally considered that treatment should start within 5 days after the onset of palsy. Most authors consider that treatment should last for a maximum of 10 days (18 days for ARIT). This means that patients can be treated for up to 4 weeks after the onset of palsy. The majority of trials (64, 65, 67) require that treatment should start as early as possible to prevent damage of the nerve; however, Sittel et al. (69) instead used a later start but a longer treatment period. It should be stressed that the trial of Sittel et al. did not use double blinding with placebo and that it was a retrospective study with a drop-out rate of almost 30% and a high proportion (72%) of patients with incomplete paresis.

Conclusions

A review of the literature of the last century regarding the treatment of peripheral facial nerve paresis reveals three major problems:

1. The etiology of Bell's palsy is unclear and, consequently, treatment options vary widely.
2. The spontaneous course of Bell's palsy has not been systematically examined, so that it has been impossible to compare the effect of treatment with the outcome of the natural history of the palsy.
3. Trials designed to prove the efficacy of a given treatment are inadequate and do not fulfill the requirements for a rigorous clinical study.

Although thermal methods, massage and facial exercises are appreciated to a very large extent by the patients, no controlled clinical trials have been published. These therapies may be considered to be forms of psychotherapy. Electrotherapy is not useful; intensive electrical stimulation may increase the risk of contractures and should be abandoned. Vasodilators are of doubtful value and no documentation of their effect exists. In terms of surgery, there is no documentation that any kind of surgery proves a better outcome for patients with idiopathic peripheral facial nerve palsies. No significant efficacy could be demon-

strated with either prednisone or combined prednisone + acyclovir treatment. Combined therapy with ARIT does not improve the final outcome.

DISCUSSION

Etiology

The aim of this study, as mentioned above, is not to discuss all possible theories but instead to describe the natural history of Bell's palsy. For reviews, see Miehlke (14) and May (21). The original cold hypothesis has been improved to an edema theory, which indicated the necessity of surgery. However, is the edema essential? It is in no way specific and can, in humans, be caused by many insults. Trauma, burns, infections, bacteria, viruses and irradiation can also cause edema in tissue. The edema is a result of damage to the normal tissue structure and is not a disease. The temporal bone contains a narrow canal with the facial nerve inside and pressure from the edema will damage the nerve. However, the edema is a secondary effect caused by several known and unknown factors, which should be considered as the primary effect. To date no proof has been given of the importance or consequence of the edema. During parotidectomia the facial nerve swelled to twice its normal diameter as a result of manipulation in difficult cases; however, no paresis developed. Steroids have been given to counteract the inflammatory edema, but their effect has not yet been proven.

Peripheral facial nerve paresis occurs in patients with many different diseases and can also be caused by infections, such as herpes zoster (21). Some idiopathic palsies may resemble an infectious disease (70) or a reactivation of HSV-1 (27) or other viruses, such as Epstein–Barr virus or cytomegalovirus (71). Recently, Vrabec and Payne (72), using a PCR assay, detected HSV and varicella-zoster virus in 42% and 44%, respectively of cases of cranial nerve ganglia. The trigeminal, geniculate, vestibular, spinal and vagal ganglia were examined. Vrabec and Payne concluded that "In order to confirm a viral etiology for various cranial nerve disorders, demonstration of a significant difference in prevalence of the viruses in specimens from afflicted individuals will be necessary". This is a logical and thought-provoking conclusion; however, a final conclusion has yet to be drawn.

Natural history

The aim of this study was to describe the spontaneous course of peripheral facial nerve palsies of unknown etiology. In addition to exposing the recovery profile of Bell's palsy, insight was also gained into many other aspects of the disease. One of the most important was that in order to draw statistically significant conclusions one requires a large number of patients; as a result the number of sophisticated statistical calculations required can automatically be reduced. When 500 patients were collected no significant conclusions could be drawn for the subgroups. When the number of patients reached 750 the situation looked better, but in some subgroups there was still no significance. Having collecting 1,000 patients the conclusions were significant and did not change after reaching 1,700 patients. The foolproof conclusion that can be drawn is that studies involving only a small number of patients are useless.

Incidence

Investigators from the Mayo Clinic, Rochester, MN have performed very careful analyses regarding the incidence of Bell's palsy: Hauser et al. (73), in 1971, found the incidence to be 22 and Katusic et al. (74), in 1986, found an incidence of 25; however, the patient samples in these two studies were small. Adour et al. (75), in 1978, found an incidence of 17–19 based on a sample of 1,048 patients. Devriese et al. (76), in 1990, based on a survey of $\approx$1,000 patients, found the incidence to be 20. In the present study the incidence was estimated to be 32. I do not have definitive knowledge of geographical and demographic differences in the incidence of Bell's palsy and therefore further investigations must be considered necessary in order to solve this problem.

Recurring and familial Bell's palsy

This sample includes 6.8% of Bell's palsy patients who had previously suffered from facial palsy on the same or opposite side of the face. Ghiora and Winter (45), in a review of the literature between 1939 and 1960, reported recurrence rates of 4.5–15%. Park and Watkins (48) found a 7% recurrence rate in a sample of 440 patients. Adour et al. (75) described previous paralysis in 9.3% of a group of 1,000 patients. Devriese et al. (76) found an 8.6% recurrence rate in a sample of 1,235 patients.

Several case reports of familial Bell's palsy have appeared in the literature, e.g. Knudstrup (77). De Santo and Schubert (78) observed 10 cases in a family and Willbrand et al. (79) reported 29 cases in a single family. The familial incidence of Bell's palsy in this study was 4.1% and in 1 family 18 cases were observed over 3 generations during a 58-year period.

Both the recurrence rate and familial incidence of Bell's palsy are surprisingly high. Genetics may play a role, but knowledge of which factors are actually inherited remains unknown. The association between human leukocyte antigen and Bell's palsy is still unclear (80). Further genetic studies are needed to resolve these problems.

Seasonal variation and clustering

No seasonal variation or clustering was evident in this sample and furthermore no variation from year to year or from one decade to another could be demonstrated (Fig. 1). Adour et al. (75) did not observe significant differences in the number of cases occurring during the cold and warm seasons and neither did they report any epidemic incidence. Devriese et al. (76) reported that far more cases of Bell's palsy were seen during the last 3 months of the year. Parry and King (81), in a 25-year study including 516 patients, found a slight, but non-significant, tendency for the condition to be more common during the winter months. Park and Watkins (48) found no statistically significant seasonal variation in a sample of 500 cases. Leibowitz (82) analyzed the time distribution of 499 consecutive cases of Bell's palsy and found it to be statistically different from the distribution of independent cases, because the cases appeared in clusters. The explanation for the finding of seasonal variation or clustering in some samples could be that the number of patients was too small. The lack of seasonal variation must be noted but exerts no influence on the etiology, even if it definitely eliminates any possibility of epidemic viral infections.

Sex distribution

The question of a gender predominance among patients afflicted with idiopathic facial palsy has been discussed in the literature, without any conclusions being drawn. One of the main reasons for the discussion has again undoubtedly been the small amount of material available. This investigation included 1,701 patients with Bell's palsy, 818 of whom were male and 883 female, a gender ratio equal to that in the general population. In almost all trials no comparison has been made between the sex distributions in the patients and in the general population. Adour et al. (75) found the proportions of males and females with Bell's palsy to be about equal. Devriese et al. (76) reported more men than women in their sample but the difference was not significant and no comparison was made with the general population. In contrast, Park and Watkins (48) found palsy to be more frequent in women than in men; however, again the sex distribution was not compared to that in the general population and the study included only 450 patients.

Side of the face

There is no disagreement in the literature about this aspect of Bell's palsy. The largest patient samples (75, 76, this study) show no significant difference in the number of palsies of the right and left sides of the face.

Age distribution

This study showed that the incidence of Bell's palsy reaches a maximum between the ages of 15 and 45 years and that the disease is significantly less common below the age of 15 years and above 60 years of age. It should be stressed that it is very important to compare the age distribution of the patients with that of the underlying population, because the different age groups are not of equal size. In spite of this, the largest series (48, 63, 75, 76) showed the highest frequencies of Bell's palsy in the second, third and fourth decades.

Time of beginning recovery and final outcome

The number of days between the onset of paresis and the beginning of remission of function is a very crucial time, during which the fate of the nerve is determined (Fig. 7). In this study, 94% of patients with incomplete paresis experienced full recovery. Of those who showed signs of remission in the first and second weeks, 88% and 83%, respectively experienced full recovery. There was no significant difference in outcome between patients who showed signs of remission in the first and second weeks, but remission in the third week was associated with a significantly worse outcome (61%). After 3 weeks a period without remission occurred which lasted until ≈ 3 months after the onset of the palsy and this is referred to as "the hibernation of the facial nerve". These patients underwent total degeneration and did not regain normal function, as previously stressed by Taverner (68) in 1965.

Ramsey et al. (66) concluded that the literature clearly shows that virtually all patients with clinically incomplete paresis have excellent recovery of facial function, independent of treatment: in this study a rate of 94% was found, Parry and King (81) found a rate of 96% and Sittel et al. (69) a rate of 98%. In the group with initially complete Bell's palsy in this study, 61% regained normal facial muscle function. In contrast, Ramsey et al. (66) did not "expect" to observe full recovery in $>40\%$ of the patients with paralysis. Steroid treatment was "expected" to improve the recovery rate to $\approx 57\%$. Therefore there seems to be no difference between the percentage of recovery observed in the present study and in the steroid-treated patients.

Age-dependent recovery

Age is another parameter that influences the final result (Fig. 8). Children have the most favorable prognosis, with 90% experiencing full recovery. Above the age of 60 years, only about one-third of patients will experience the return of normal function. Adour and Wingerd (83) described a worse

outcome for patients aged >60 years. Prescott (63) and Devriese et al. (76) emphasized that the age of the patients was an important factor determining recovery. Katusic et al. (74) reported that those aged ≥ 55 years had a significantly higher rate of incomplete recovery. The influence of age on the final outcome is therefore highly significant.

Postauricular pains

In this study, postauricular pains were registered in 52% of all Bell's palsy patients. Patients with pains have a significantly worse prognosis than those without pains. Katusic et al. (74) evaluated prognostic factors and reported that pain other than in the ear had a significant relationship with incomplete recovery.

The prognostic value of topographical tests

Figure 9 shows the prognostic value of the three topographical tests: taste, stapedius reflex and nasolacrimal reflex. In the literature a dry eye has always been associated with a bad final outcome. Adour and Wingerd (83), May et al. (60) and Katusic et al. (74) found a worse outcome in patients with dry eyes.

Hypertension

The influence of hypertension on the outcome of Bell's palsy is controversial. Adour and Wingerd (83), in a sample of 446 patients with Bell's palsy, found 55 (12%) with hypertension and demonstrated a significantly higher risk of a worse outcome in these patients. Devriese et al. (76), in a sample of 1,235 patients with Bell's palsy, diagnosed 145 patients (13%) with hypertension. They attributed the worse outcome for the patients with hypertension to the higher age of those patients.

In this study, 134/1,701 patients (8%) suffered from hypertension. A total of 264 patients were aged >60 years and 37 of these patients (14%) had hypertension. Bell's palsy did not occur at a higher frequency than expected in patients with hypertension. The frequency of sequelae in patients with hypertension did not differ from that in the group with normal blood pressure. Katusic et al. (74) confirmed the hypothesis that hypertension is a risk factor for Bell's palsy and also concluded that increasing age resulted in a greater frequency of incomplete recovery. The importance of comparing hypertensive patients with non-hypertensive patients of equal age should be emphasized.

This study (Fig. 8) shows that recovery depends to a very large extent on age, as also stressed by Devriese et al. (76), so the connection between advanced age and hypertension would seem to be a coincidence. Another explanation for this finding may be misdiagnosis of an incomplete central paresis as a peripheral paresis.

Sequelae

The factors determining the degree of sequelae are paresis, contracture and associated movements (Tables IX and X). The mechanism that causes contracture is not known in detail, but some degree of regeneration is necessary. Contracture is almost always combined with associated movements or synkinesis caused by misdirection of the nerve fibers during regeneration. It is necessary to follow these patients for 1 year, until a steady state is attained.

In this study, associated movements and contracture were found in 16% and 17% of patients, respectively. Adour and co-workers (59, 67) found contracture and synkinesis in 15% of prednisone-treated patients and 21% of untreated patients, respectively in 1972 and in 28% of prednisone (placebo)-treated patients and 13% of acyclovir + prednisone-treated patients, respectively in 1996. Austin et al. (64) described synkinesis in 13% of both steroid-treated and untreated patients. Therefore, prednisone does not seem to prevent sequelae.

How to avoid misdiagnosis of Bell's palsy?

Although Bell's palsy is the commonest form of peripheral nerve palsy, care should be taken not to misdiagnose any palsy as idiopathic. Although some workers try to determine the etiology by using a wide range of tests, this approach is not always successful. The symptomatology of Bell's palsy is not specific and if the acute phase of the palsy does not affect all branches it is important to consider another etiology because a partial palsy of one or two branches strongly suggests disease localized distally from the stylomastoid foramen, and may indicate a cancer in the parotid gland.

If symptoms from other cranial nerves are observed an obvious supposition would be viral infection or neoplastic disease. In 75% of cases a peripheral facial palsy caused by herpes zoster shows acoustic and/or vestibular disturbances. The diagnosis can be verified by CSF or serological tests if typical vesicles are not seen. If endocrinopathic diseases are suspected, diabetes mellitus is a possibility and can be verified by a glucose tolerance test. If the parotid gland is enlarged and eye symptoms are observed, sarcoidosis is a possibility and can be verified by lung X-ray and possibly by mediastinoscopy with gland biopsy. A recurrent palsy should be examined using standard investigations and, in addition, eye and neurological examinations should be performed. CT scans of the facial nerve canal should be made even if pathology seldom occurs. Of 69 patients

with recurrent Bell's palsy in this study, only one had a pathology of the facial nerve canal, a bifid canal answering to the third part which shows that this type of pathology is very rare.

Among 13 other patients with recurrent palsy of different etiologies 1 patient with ear cholesteatoma was found. Another had pronounced destruction of the second part of the bony canal involving the geniculate ganglion region. Upon surgery a tumor was found and histology revealed the same picture as that of a breast cancer for which the patient had been treated 27 years earlier. Another patient with radiologically demonstrable destruction of the temporal bone suffered from Paget's disease.

As mentioned before, Bell's palsy is an acute, monosymptomatic, peripheral facial palsy of unknown etiology. A monosymptomatic diagnosis is required in order to distinguish the paresis from tick-borne B. burghdorferi infection, collagenosis and polyneuritis, for which the symptoms are polysymptomatic. Specific laboratory tests are required in order to differentiate between many of these diseases.

Table II shows the timetable for examinations of peripheral facial nerve palsies. Using a follow-up timetable with short intervals between visits provides many chances for assessing and reassessing the diagnosis. Furthermore, supplementary tests can also be performed. In this study the preliminary diagnosis was changed during follow-up in $\approx 2\%$ of all patients.

Side-effects

Surprisingly, very few of the publications that have tried to prove the effect of steroids have revealed the frequency and severity of side-effects. Elderly patients are excluded from many trials because the treatment could cause severe side-effects. It is obvious that the results of such studies would be over-exaggerated, because age is a very important factor determining recovery (63, 74, 76, 83; Fig. 8).

A drug as potent as prednisone, given in high doses, is estimated to produce side-effects in $\approx 10\%$ of patients. Adour et al. (59) used one of these side-effects as a indication for treatment: the treated patients were euphoric and the untreated patients asked for treatment.

Psychotherapy

All patients with acute peripheral facial nerve palsy complain of anxiety. The best way to help these patients is to explain that the symptoms are caused not by a stroke but by a lesion of the peripheral facial nerve, maybe of unknown etiology. The patient should be informed that 85% of patients begin recovery within 2–3 weeks and that in the other 15% beginning function is obtained within 3–5 months. All patients are tested according to the scheme shown in Table IV. Patients should be told that 3/4 patients achieve full recovery with normal mimical facial function within 3–4 months.

Follow-up of patients should occur once a week initially. It is essential that patients feel better, are less depressed after the consultation and are looking forward to beginning recovery. Many patients feel comfortable with moderate heat applied to the paretic side of face. Facial exercises have a great psychological effect: when function starts to return the patient can see the improvement. In my experience giving the patient information and repeating follow-up is the best way to help them; however, it should be stressed that there is no documentation of the efficacy of any kind of treatment.

Ethics

The Third International Nerve Symposium was held in Zürich in 1976. The Swedish professor Hamberger (84) participated in a panel discussion and stressed the importance of thorough examinations, frequent follow-up and the provision of detailed information for patients with peripheral facial nerve palsies. No treatment was given by Hamberger with the exception of psychotherapy, which included mild physiotherapy. This seems to me to be a honest way to manage patients with peripheral facial palsies. Nevertheless, Katusic et al. (74) from the Mayo Clinic expressed in a publication from 1986 that "The extreme attitude, according to Hamberger, is that of the Scandinavian physicians who only treat patients with Bell's palsy with physiotherapy or massage as a 'psychological treatment' " and continued: "Our results suggest that individual differences in patients may influence the type of therapy chosen". A close reading of the publication revealed a retrospective study of a small sample without randomization. The trial was not double-blind or placebo-controlled and no indication was given as to how individual differences in patients may influence the type of therapy. Nevertheless, the authors recommended treatment with prednisone. Using high-dose prednisone is risky and, without documentation of the final outcome, may be unethical.

It is time, after two to three decades, to require valid documentation before using prednisone. This raises an ethical problem: how can such a high-potency drug with marked side-effects have been used at high doses for such a long time without documentation? It is unbelievable and must raise a huge ethical dilemma for those doctors who prescribe prednisone for Bell's palsy patients. Now we are back to the accusation of Hamberger (84) by Katusic et al. (74). Hamberger does not have an ethical problem because he was honest. Katusic et al. do not have any basis

for their accusation and treatment; this is an ethical problem of large dimensions and they are still using the drug without proof of its efficacy and whilst only informing the patients of the benefits. Even worse is the fact that the euphoric side-effect is used as an indication for the use of the drug (59), which is very unethical.

Final remarks

Evaluation of therapy for Bell's palsy is made difficult by the spontaneous and usually complete recovery observed in the majority of patients. In this study, 71% of all patients regained normal facial muscle function.

In a trial of treatment only $\approx 20\%$ of patients need the treatment, meaning that a large number of patients are required in order for the trial to be valid. According to Burgess et al. (61) at least 200 patients are required in both the treated and placebo groups. The most striking flaw of many trials is the insufficient number of patients enrolled. In many trials the aim seems to be to show the effect of a given treatment rather than to make an objective evaluation of the data and a statistical analysis. It seems obvious that bias is unavoidable (85). Furthermore, discrepancies are often found between the results of the data analyzed and the conclusions. Careful reading is crucial.

As mentioned above, no documentation of the efficacy of physiotherapy, electrotherapy or surgery has been given. In a review of the literature in 1987, Stankievicz (62) concluded that a definitive and statistically valid study considering the benefit of steroids in the treatment of Bell's palsy had yet to be performed. Even today the situation remains unchanged, because no valid study to prove the effectiveness of predisone has been published.

Maybe it is time to investigate new directions in the treatment of Bell's palsy. A parallel can be drawn with Guillain–Barré syndrome (86), an acute immune-mediated polyradiculoneuropathy preceded by an infection with Campylobacter jejuni, mycoplasma, cytomegalovirus or Epstein–Barr virus. Today's treatment is i.v. immunoglobulin in high doses (87). Steroids do not help these patients, but were used for many years.

It would be most logical to first prove the etiology of Bell's palsy, as it is obvious that the best treatment for a disease can be given when its etiology is known. Bell's palsy is not a disease sui generis but a paresis caused by several known and unknown generating factors. When is the optimal time to start treatment? Generally, all kinds of treatment should be started as soon as possible, as long as the diagnosis is confirmed. The electrophysiological examinations performed by Langworth and Taverner (32), Olsen (34) and Esslen (35) showed that degeneration occurred within 7–10 days. Based on these results, the tendency was to start treatment, either surgery or prednisone treatment, as early as possible. Pulec (88, 89) advocated early decompression. Brown (90), Austin et al. (64) and Shafshak et al. (65) recommended early prednisone treatment. As stressed by Adour et al. (67), no electrical test is able to predict the outcome in all patients but ENoG seems to be the most valuable (91). When Wallerian degeneration has started, it takes up to 3 days before the nerve is inexcitable; this means that one is always about 3 days behind the progression of degeneration. The first week seems to be the crucial time for the nerve to survive and if any treatment could help it is obvious that it should be started as early as possible.

It is recommended that prospective investigators should read the paper by Schulz et al. (85). The conclusion is "This study provides empirical evidence that inadequate methodological approaches in controlled trials, particularly those representing poor allocation concealment, are associated with bias. Readers of trial reports should be wary of these pitfalls, and investigators must improve their design, execution, and reporting of trials". The tutorials by Neely and co-workers (92–95) are also highly recommended.

CONCLUSIONS

The Copenhagen Facial Nerve Study collected a large amount of data and, based on an analysis of these data, the following statistically significant conclusions can be drawn.

- Time of beginning recovery of function: early recovery leads to excellent function and late recovery implies sequelae.
- Age of the patient: young patients have a good prognosis and elderly patients a worse outcome.
- Topographical tests: a dry eye and abolished taste and stapedius reflex affect the prognosis negatively.
- Postauricular pains: patients without pains fare significantly better than those with pains.
- The incidence of Bell's palsy is estimated to be 32 per 100,000 population per year.
- Recurrent palsies were found in 7% of Bell's palsy patients and 4% of all cases of Bell's palsy represented familial Bell's palsy.
- Seasonal variation and clustering were not found and variations from year to year or from decade to decade could not be demonstrated because the influx of Bell's palsy patients remained constant over the 25-year period of the study.

- The investigation clearly indicates that there is no difference in the sex distribution of Bell's palsy patients.
- There is no difference regarding left or right localization of Bell's palsy.
- The age distribution of Bell's palsy patients should be compared with that of the underlying population. The incidence reaches a maximum between the ages of 15 and 45 years. It is significantly less common below the age of 15 years and above the age of 60 years.
- Complete recovery was observed in 71% of all patients. Males and females recovered in the same proportions.
- Patients with incomplete paresis regained normal function in 94% of cases and those with complete paralysis regained normal function in 61% of cases.
- All Bell's palsy patients recovered to some extent: 83% of all patients recovered with a fair result; 5% of patients were left with an unacceptably high degree of sequelae.
- Normal function was regained within 3 months in about two-thirds of all patients; after 6 months no additional patients regained normal mimical function.
- Sequelae were found as follows: residual paresis, 29%; contracture, 17%; associated movements or synkinesis, 16%; dry eye and crocodile tears, 2%.
- Bell's palsy is not more frequent in hypertensive patients and the prognosis in these patients is not worse than that in non-hypertensive patients; the connection between advanced age and hypertension would seem to be a coincidence.
- Pregnant women with peripheral facial nerve palsy regained normal function in 61% of cases.
- Paresis caused by herpes zoster was complete in 88% of cases and incomplete in 12%. The prognosis for these patients is bad, with normal recovery in 21% and no recovery in 4%.
- Facial nerve palsy in diabetics was incomplete in 62% of cases and complete in 38%. In spite of the high proportion of mild pareses the final outcome, as a result of polyneuropathy, was poor, with only 25% regaining normal function.
- Bell's palsy in children is uncommon and the prognosis is excellent, with 90% regaining normal function. The non-Bell's palsy group had different etiologies determining the prognosis. The incidence of birth trauma facial nerve paresis has decreased dramatically during the last 25 years, as a result of improvements in obstetrical techniques.
- Bell's palsy has the best prognosis of all peripheral facial nerve palsies.
- Based on the experience of this investigation it should be emphasized that clinical trials require a large number of patients in order to be valid.

The facial nerve surgeon from London Sir Terence Cawthorne advised his assistants to examine facial nerve patients properly. It would be a catastrophe to diagnose facial palsy as idiopathic if the patient suffered from a severe disease:

"All that glitters is not gold" (William Shakespeare)

"All that palsies is not Bell" (Cawthorne)

For more than 35 years I have studied facial nerve paralysis and have learned that close reading of the literature is crucial:

"To be or not to be" (William Shakespeare)

"To be skeptical" (Peitersen)

FUTURE DIRECTIONS

Regarding idiopathic peripheral facial nerve paralysis: where are we and where are we going? The focus should be on the treatment. Today's problems are similar to those encountered in the 1950s and 1960s, when facial nerve surgeons believed in decompression operations but never provided significant documentation of its efficacy. Prednisone treatment began in the 1970s and increased dramatically in the 1980s and 1990s, especially in the USA but less so Europe and has never really been accepted in Scandinavia. A survey of the literature today reveals at least 100 trials that have reported the effectiveness of prednisone treatment. Nevertheless no trial has fulfilled the requirement for a clinical, randomized, double-blind study with a sufficient number of patients and with exact descriptions of the time of recovery and sequelae, including residual paresis, associated movements and contracture, graded after 1 year. This is a very depressing statement to make but nevertheless it is the truth.

Simple electrical tests have been used to predict the degree of denervation. Based on these results the final outcome was predicted, but this was not based on a clinical examination after 1 year. The value of electrical tests has been overestimated. If the nerve is inexcitable then Wallerian degeneration must have begun 3 days previously and the denervation has to be accepted. There is no test available that can tell us early enough what has happened to the nerve. From this point of view any viable treatment should be begun as early as possible. But is there a drug of choice? Not yet is the correct answer. Early treatment raises another problem: $\approx 80\%$ of Bell's palsy patients do not need treatment and it may be unethical to treat this group, given the chance of side-effects.

The enormous use of prednisone over the last two decades is a huge problem. To use a potent drug with a high risk of side-effects, but without documented efficacy, is unethical. Although patients benefit from the euphoric side-effect they do not have the chance to differentiate between the effect and the side-effect. In my opinion this seems to represent a real ethical problem for doctors. The time has come to stop the use or misuse of prednisone, to be honest with the patients and to accept that there is no effective treatment for Bell's palsy.

Future etiologic studies should be expanded to reveal the pathogenesis of Bell's palsy. Obtaining additional knowledge may open the door for a specific treatment. Last but not least, how can the effect of all kinds of treatment be explained? Only by following the spontaneous course of idiopathic peripheral facial nerve palsy.

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