The facial tics

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Facial movements of a stereotyped pattern are not infrequently seen in neurological, ophthalmological and neuro-otological practice. These movements are similar to coordinated movements, often with rythmic intervals, but usually occur in non-rythmic sequences; they simulate voluntary movements but are involuntary and usually affect several muscles or groups of muscles; and, starting around the eyes or mouth they may spread even as far as the shoulders. As do most extrapyramidal automatisms these facial movements tend to disappear during sleep. Voluntary suppression whilst possible for brief intervals is followed by a marked feeling of tension. There is probably an organic basis in the genesis of all tics.

The facial tics may be classified as:

1. *Functional tics* without known organic basis and usually bilateral.

2. *Tics whose organic basis can be shown* to be localised in the extrapyramidal system, such as post-encephalitic, drug-induced and the striatal tics. The drug-induced tics occur in patients receiving large doses of levodopa for the treatment of Parkinsonism, or the anti-emetic metoclopramide. Tardive dyskinesia is seen occasionally after the use of phenothiazine preparations, though usually after prolonged treatment in high dosage, but the movements may then remain permanently even after complete cessation of the therapy.

3. *Tics whose organic basis is seldom proved*, usually bilateral and typified in Gillies de la Tourette's generalised tic disease.

4. *Tics resembling the functional tics* superficially but are rarely bilateral and if so are not synchronous on the two sides either in intensity or in strength. The movement is initiated by the peripheral motor neurone and varies from fibrillation and lightning shock-like movements to severe spasms which may become tonic or even tetanic. The shoulder and the neck are seldom involved.

This latter group may be subdivided:

a. *Tics associated with neurological disorders*, which can usually be distinguished by accompanying neurological signs.

b. *Isolated neurological or psychological phenomena* in which an electromyographic abnormality can be demonstrated.

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This sub-group should again be divided into:

I. *Tics associated with cortical irritation* are rare and the diagnosis may usually be made with the help of an electroencephalogram. Epilepsy partialis continua may produce a picture very similar to hemifacial spasm, but the the spread of the contraction to the arm and hand facilitates the diagnosis.

II. *Psychogenic conversion reactions* (habit spasm) in which the electroencephalogram is normal and the movement is abolished by sodium amytal injection.

III. *Lesions at or below the 7th nerve nucleus* which may be cryptogenic such as infections or degenerations, lesions of the peripheral 7th nerve or post-paralytic.

The neuro-otologist is primarily concerned with the last subsection of the groups of tics, usually termed hemifacial spasm. Isolated facial tics involving one or two branches of the facial nerve may anticipate by several years a hemifacial spasm, or remain selective, when the buccal branch is most often involved. Despite the epidemic of encephalitis lethargica which left many cases of facial spasm in its wake it is still widely held that the condition is of psychogenic nature.

Clinical findings

The spasms last a few seconds to 15 minutes, the individual twitch being of half to 60 seconds in duration. The spasms usually though not always disappear during sleep, but the patient may be awakened by the spasm. The condition is very unusual in children and most authorities agree that it occurs most frequently in women.

Spasm is *triggered* by emotional or by motor mechanisms such as forcible eye closure. Paradoxical movements such as spasmodic lifting of the corner of the mouth upon closure of the eyes may occur with a latency of some 5 seconds. The movements can only be repressed in certain cases with great difficulty and can seldom be repeated voluntarily. It is unusual for patients with spasm to show obsessional impulses or psychopathic signs.

The *onset* is insidious and subacute, complete spontaneous disappearance is rare. The condition continues with intermissions and exacerbations.

Blepharospasm is a localised spasm of the muscles of the eyelids and is almost always bilateral. Often the lids are so firmly pressed together that the patient is functionally blind for as much as one third of this wakening hours, rendering most occupations and even household duties virtually impossible. Though other muscles may be involved eventually, in them the spasm is seldom severe.

Tic convulsive is the term given by Cushing (1920) to the facial spasm associated with trigeminal neuralgia in which he found a single lesion

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affecting both 5th and 7th nerve roots. This may be a vascular abnormality or a condition such as an epidermoid. The pain in such a case of course disappears after blocking of the 5th nerve.

The finding of contractions of the stapedius muscle simultaneous with the contractions of the facial muscles (Diamant *et al.*, 1967) which was noted in three of the 10 cases in which it was sought in the present series, suggests that the lesion may be situated in the facial nerve proximal to the nerve to the stapedius muscle.

Diagnosis

The diagnosis of the individual forms of tic is usually not difficult bearing in mind the characteristics already described under classification of these movements.

Electromyography may be used to differentiate certain of the tics in group 4. Spontaneous activity is absent in hemifacial spasm yet in post-paralytic reflex tic a continuous activity occurs at rest with a pattern similar to that of myotonia. During the spasm the rhythmic impulse trains differentiate the condition from a central process and suggest that the basis of the lesion is in the peripheral nerve.

Mastoid and petrous X-rays and a full neuro-otological examination are advisable to try to exclude lesions irritating the 7th nerve.

The functional tic is repetitive, controllable, monotonous, a natural movement which has become a compulsion in a patient with a psychopathic predisposition.

An isolated epileptic focus near the cortical facial centre may produce attacks of facial spasm with a regular pattern, usually affecting the lower face, which are shorter and usually more violent than the typical hemifacial spasm. Neighbouring cortical centres are eventually affected.

Fasciculations usually occur only in paralyses associated with lesions of the peripheral pathways and are an expression of a process of irritation and slow destruction of the root cells of the anterior horns. They occur in a variety of diseases which slowly affect the anterior horn cells as painless synchronous contractions of all the fibres of a motor unit, shown as brief rapid contractions of the muscle fibres, which are seen under the skin but do not move the part to which the muscle is attached. Brief physiological fasciculations occur in response to cold especially when the subject is fatigued.

The benign form of *myokymia* is the commonest type of facial movement and usually affects the orbicularis oculi, especially the lower lid, at times of strain or fatigue. *True facial myokymia* is rare and appears as an irregular writhing continuous movement of the side of the face, the muscles being in a constant if slight contraction with narrowing of the palpebral fissure and raising of the angle of the mouth. The movement is absent during sleep and unassociated with voluntary contractions, it may last up

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to 6 months before it tapers off. The condition is caused by a lesion in the medulla close to the facial nucleus usually in disseminated sclerosis of which it may be the first sign, and rarely in such conditions as brain stem tumours when it is often accompanied by facial weakness.

Treatment

Medical

The disability caused by the irregular spasm frequently causes the patient much distress. In severe cases the medical treatment is seldom of long term help and if persisted with serves further to undermine the patient's confidence and general health. Centrally acting drugs such as Artane which lessen the motor impulses to the voluntary muscles have not proved of value. Similarly on the suggestion that the condition may be based on vascular spasm vasodilators and intravenous procaine have been tried but have not been shown to be of value, nor have psychotherapeutic drugs, belladonna alkaloids and related medicaments and tranquilising drugs. Carbamazepine or phenytoin help in some cases of mild hemifacial spasm, also in some milder cases dark glasses may be of help by partly concealing the movements and by lessening the effect of light, which is frequently a precipitating cause.

Indications for operation. For successful treatment it should be emphasized that the patient should desire to be cured and no patient should be persuaded against his will to have treatment, especially surgical, for any form of facial spasm. It may be desirable to demonstrate to the patient the facial palsy which he is likely to have for at least a time after operation. This may be done by paralysing the facial movements temporarily with local anaesthetic placed just lateral to the styloid process. If he is then allowed to discuss the palsy at home with his family he should be better able to understand one of the major consequences of surgery.

Surgical treatment

Before considering local surgical treatment of hemifacial spasm intracranial causes of the condition must be excluded. Many reports have appeared of recurrent spasm secondary to vascular lesions in the cerebellopontine angle, cirsoid aneurism of the basilar artery, arterio-venous malformation or loop of the antero-inferior cerebellar artery (Janetta, 1972). Surgical treatment in this area involves the danger of damage to the neighbouring structures and often it is difficult to maintain the separation between nerve and vessel. Facial twitching has long been recognized as an early manifestation of an acoustic neuroma and two such cases occurred in the present series.

The frustrations of the treatment of facial spasm are evidenced by the numerous approaches to the problem in medical literature. Partial or complete cessation of the spasm for longer or shorter periods of time may

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be obtained by injury of the nerve in the parotid gland, in the third part of the nerve in the parotid gland, in the third part of its course in the temporal bone or in its course above the middle ear where it is easily viewed. The form the injury takes varies with the individual surgeon, the nerve may be crushed, needled with plain or diathermy needle (Waksugi, 1972; Ludman, 1973) or it may be hemisected and the proximal cut half of the nerve sutured back to the nerve trunk (German, 1942; Scoville, 1955; Mielke, 1961; Diamant, 1967). This latter form of treatment which has received considerable support reduces the number of nerve fibres in the peripheral part of the nerve. In the present series three early cases were treated by a technique similar to that of Scoville (1955), all showed prolongation of the resulting facial palsy and return of the spasm, though this was less marked than previously.

The simplest surgical approach is to inject a local anaesthetic, alcohol or a sclerosing agent in the region of the facial nerve as it leaves the stylo-mastoid foramen, or, by using electromyography to localize the site of the branch of the nerve to the affected muscle, this branch alone may be injected (Blumenthal, 1972; Totsuka *et al.*, 1972). Relief was obtained for up to two or three months by injection of alcohol close to the main nerve trunk in four cases early in the present series, usually at the cost of facial palsy, but the injection is difficult, painful and unpredictable, particularly on repetition, so that the patient's confidence is soon undermined. The spasm may be aggravated in due course by the intraneural injury.

Decompression of the nerve was suggested on the thesis that the lesion was in the facial canal in the temporal bone, initially it was thought to be in the third division proximal to the stylo-mastoid foramen (Woltman *et al.*, 1951; Cawthorne and Haynes, 1965; Curtin, 1969) later as neuro-otological surgery developed decompression of the entire course of the 7th nerve in the temporal bone was tried (Pulec, 1972). Abnormalities such as oedema, fibrous constriction of the perineurium or diffuse thickening of the nerve have been described. Celis-Blaubach (1974) voices the generally held view that the results of this procedure depend upon the extent of the surgical injury to the nerve, the more severe and long lasting the facial palsy post-operatively the better the results.

Permanent cure in a reasonable proportion of cases with hemifacial spasm can only be obtained by complete division of the nerve to the affected muscles and the prevention of the re-establishment of nerve continuity or by effective myomectomy of the affected muscles alone or combined with nerve resection, a procedure which is difficult, results in facial scarring and does not always give total relief of the spasm, either immediate or long term.

Early workers (Greenwood, 1940; German, 1942) and since then a number of authors (Finochietto and Marino, 1950; Harvey, 1971; McCabe, 1970 to 1972; Potter, 1972; Nosik and Weil, 1956; Mielke, 1961; Diamant

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and Enfors, 1967; Reynolds *et al.*, 1967) have suggested *selective neurotomy*, selective crushing of (Sjoquist, 1967), or evulsion of the nerve branches to the individual muscles. These procedures are very effective especially in blepharospasm, but, owing to post-operative fibrosis, are difficult to repeat if nerve regrowth takes placed (Diamant and Enfors, 1967). In many cases also paralysis of the muscles supplied by the divided nerves is troublesome to the patient and complications such as ectropion, keratitis and epiphora may occur. The communications between the different branches, both main and secondary, of the 7th nerve are so numerous that it is difficult at times to divide them all and if the nerve be divided proximal to these communications regrowth of the nerves to the affected muscles is followed by increasing trouble with the spasm. The procedure is more effective if carried out distally for a more localized condition such as blepharospasm (McCabe, 1970). In the cases of very severe and widespread hemifacial spasm the condition is so extensive that operations such as these are not always practical and it has been considered advisable to section the 7th nerve trunk closely proximal to its division in the parotid gland and rather than to leave the patient with a permanent facial palsy the distal cut end has been anastomosed to the hypoglossal nerve (Harrison, 1973). To emphasize the facility with which return of 7th nerve function and of spasm may take place even after complete nerve section, though in the first two cases the facio-hypoglossal anastomosis was successful in restoring facial movement, the spasm returned within the year owing to the regrowth of 7th nerve fibres into the anastomosis, the muscles being supplied by fibres from both 7th and 12th nerves. The recurrence of the spasm was at first puzzling, but was easily remedied by a short second operation to evulse the facial nerve fibres from the anastomosis and at the same time to ligate the proximal end of the 7th nerve with silk thread. Six further cases showed an uneventful recovery of nerve function and no return of the spasm.

Nerve anastomosis appears first to have been described by Colemann (1937) and by Peet and Echols (1946) with notable success. It was suggested that the descending branch of the hypoglossal nerve be anastomosed to the distal segment of this latter nerve in an attempt to avoid atrophy of the affected half of the tongue, but this complication has not been a notable complaint of patients in the present series and should not be considered a drawback of the operation (Logue, 1968). The absence of emotional movements of the face is of more consequence, but all the patients were so greatly relieved to be free of the spasm that they remained fully satisfied. Hemiatrophy of the tongue has been avoided in some patients of the present author in which a facio-hypoglossal anastomosis was performed after removal of large acoustic neuromas, by obtaining a suitable length of facial nerve by dissecting out its 2nd and 3rd divisions from the temporal bone and suturing this into the hemisected 12th nerve. The distance the

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nerve fibres must then grow is much increased and the movement of the facial muscles takes considerably longer to return and hence is unlikely to be so complete as when the anastomosis is performed at or below the branching of the nerve in the parotid gland. This and the advantage given by immediate anastomosis after division of the 7th nerve accounts to a considerable degree for the better results of anastomosis in patients with facial spasm.

In patients with less severe spasm division of the whole 7th nerve is better, of course, avoided and the nerve section has in 5 patients been limited to the upper division of the nerve in the parotid gland. No difficulty has been found in obtaining sufficient length of the 12th nerve to allow an adequate anastomosis and, with so short a distance from the end plate, down-growth of the fibres and recovery of the muscle function has been rapid.

The incision to expose the hypoglossal nerve in the neck is an extension of the posterior end of the routine parotidectomy incision below and behind the lobule of the ear, it continues downwards and obliquely in the neck towards the cricoid cartilage. The two incisions give an adequate exposure of the parotid gland and the communications in the cheek between the two divisions of the facial nerve distal to the parotid gland. The facial nerve is usually exposed as it enters the posterior surface of the parotid gland using a pair of fine curved Spencer Wells artery forceps to separate the glandular tissue. The main trunk of the nerve divides into upper and lower branches soon after entering the gland. If it has been decided to retain the lower division dissection of the two divisions is carried forward through the gland into the cheek and the small communicating branches between the two divisions are evulsed from the lower division. A careful and detailed removal of all these tertiary branches must be carried out with the aid of a stimulator. The buccal branch supplying the corner of the mouth and the upper lip usually arises from the upper division, but the accompanying Figs. 1 and 2 show its anatomical variations. If it arises from the lower division it may be divided or left intact depending on the extent of the spasm. It is certainly more convenient if it adopts the commoner pattern and arises from the first division of the 7th nerve.

After the dissection of the branches is complete the upper division is tied tightly with silk distal to its origin from the main trunk and divided a little proximal to the first of its own branches. The cut distal end is then anastomosed to the 12th nerve which has been divided as far forward as possible. There should be little difficulty in obtaining sufficient length of nerve after it is freed and brought up to the parotid area through a tunnel deep to the more superficial tissues.

After the anastomosis is complete and before the wound is closed the main 7th nerve trunk should be stimulated to ensure that all connection between the upper and lower divisions has been severed, should contraction

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of any of the muscles supplied by the upper division occur an immediate revision of the anterior portion of the dissection should be undertaken. Similarly if after operation denervation is found to be incomplete revision of the procedure is indicated.

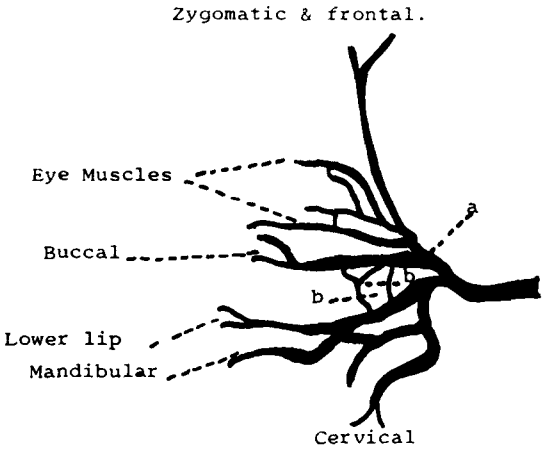


FIG. 1.

Shows the most common arrangement of the 7th nerve branches where the buccal branch arises from the upper division of the nerve soon after its origin from the main trunk. The upper division is then sectioned at (a) and the communicating branches (b) demonstrated by a stimulator and all avulsed. Facio-hypoglossal anastomosis at (a) may then be undertaken.

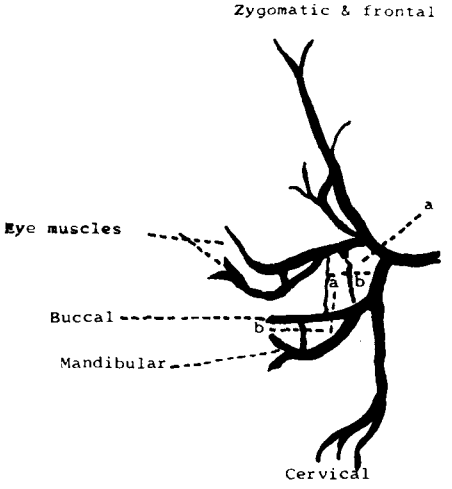


FIG. 2.

Demonstrates an arrangement of the branches of the 7th nerve frequently encountered. Here the buccal branch arises from the lower division. Section of the main nerves is carried out at (a). Communicating fibres between buccal and mandibular divisions should be sought and divided.

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In patients with bilateral blepharospasm nerve avulsion as practised by Reynolds *et al.* (1967) is carried out on both sides or facio-hypoglossal anastomosis may be used on one side and avulsion on the other. Operation on both sides is required to obtain maximum effect on either side.

Discussion

The first of the patients in this series with facial spasm was seen more than twenty five years ago and the earlier cases were followed for up to 15 years, the type of treatment changing with the experience of the various forms of surgical treatment.

Eleven of the patients seen more recently have been followed at varying intervals usually 3–6 monthly over the past 5 years. Six patients have attended for less than 5 years, the shortest for four and a half months.

Facial neuroanastomosis has proved to be a satisfactory procedure and if the suturing be carried out under magnification and with fine (8/0) silk more than 90 per cent of successful takes may be expected when the suture is performed at once after division of the nerves.

Summary

The abnormal movements of the facial muscles are classified and the diagnosis and treatment considered in light of 22 cases of hemifacial spasm.

Forms of treatment which give only temporary cessation of spasm and require complex and unpleasant treatment to be oft repeated are seldom tolerated for long by the patient.

Permanent cure of spasm can only be obtained when the facial nerve or its branch to the muscle is permanently divided or the muscle itself excised.

Neurotomy as practised by German (1942) and Scoville (1955) has not been found satisfactory in the present series.

Neuro-anastomosis at different sites in the course of the 7th nerve is discussed and shown to be of value in patients with severe widespread spasm and in those in which the spasm is more localised. This method results in only temporary palsy of the facial muscles, the duration of the palsy depending on the site of the division of the facial nerve.

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