From the Neurological Clinic
of the University Hospital Eppendorf

Department of Neurosurgery
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The spasmodic torticollis

treatise with special consideration of a new surgical therapy approach

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Contents

Abbreviations

BMLA : Bilateral microsurgical lysis (decompression) of the spinal accessory nerve roots

CT : Computed tomography

DCS : Cervical (high) dorsal spinal cord stimulation

DT : Dystonic torticollis

EMG : Electromyography

EEG : Electroencephalography

EP : Evoked potentials

MRI : Magnetic resonance imaging

PEG : Pneumencephalography

PET : Positron emission tomography

SANRs: Spinal accessory nerve roots

SCM : Sternocleidomastoid (muscle)

ST : Spasmodic torticollis

“Spasmodic torticollis can be such an unrelenting and demoralizing disability that exploration in humans of new surgical techniques requires no apology.“(T.P. MORLEY, 1976)

1.0 Introduction

Although the clinical picture has been known for a long time, there is no opinion on the etiology and genesis of spasmodic torticollis (ST). Although several different hypotheses on the pathogenesis of ST have been put forward, the cause of the disease has yet to be established. The discussion about the possible causes of this disease continues (BRISSAUD, 1895, FOERSTER, 1929, 1933, STEYERTHAL, 1906, CURSCMANN, 1907, KOLLARITS, 1908, SCHÜRMANN, 1953, BRÄUTIGAM, 1954, KRAYENBÜHL and YASARGIL, 1965, HAMBY and SCHIFFER, 1969, SVIEN u. CODY, 1969, HASSLER and DIECKMANN, 1970, SHEEHY and MARSDEN, 1976, MARSDEN and HARRISON, 1975, MUNDINGER, 1977, LOZANO-SAVEDRA, 1979, MITSCHERLICH, 1979, SIROKY et al., 1980, MARSDEN, 1980, LÜCKING, 1980, KASTE et al., 1981, van WAVEREN, 1982, THÜMLER, 1983, JACOBI, 1983, WITZMANN et al, 1984, COLBASSANI and WOOD, 1986, BRONSTEIN and RUDGE, 1987, BERTRAND, 1987 and many others). This results in the very broad spectrum of methods proposed for the treatment of ST. The treatment results are correspondingly unsatisfactory, as causal therapy is not possible if the cause of the disease is unknown.

If one traces the relevant literature on the subject of "Spasmodic Torticollis", one comes across a work by STEYERTHAL (1906) with the title: “Zur Geschichte des Torticollis spasmodicus“("History of spasmodic torticollis"). In this essay, which summarizes the previous literature at that time, the author states the following: The term “torticollis”, actually the description of a symptom, namely the “crooked” neck, comes from the French and was probably first coined by the probably first used by the French physician and humanist Rabelais (1494-1553). The first case description goes back to Felix Plater from Basel (1616). The first detailed dissertation “De capite obstipo” was written

by Georg Friedrich Jäger 1737 in Tübingen.
The term “Caput obstipum spasticum” is still used today by some authors, e.g. the anatomist LANG (1982). KRAYENBÜHL and YASARGIL (1965), on the other hand, use the term “caput obstipum” for torticollis caused by diseases of the cervical spine and its neighboring (muscles, bones, intervertebral discs, joints, ligaments, inflammation of the pharyngeal and laryngeal organs).

In German and international usage, the term “spasmodic torticollis” (ST) has generally been used and established. The term “torticollis spasticus” is used less frequently in German-speaking literature. Hereby it should be noted that earlier authors used the two terms to describe clinically different manifestations of the condition (STEYERTHAL, 1906). The term “torticollis spasticus” is used to describe the tonic form of the disease, "torticollis spasmodicus" the clonic form of the disease. In the tonic form (tonic neck muscle spasm), the head appears to be “fixed” in the incorrect position. The clonic form, on the other hand, is characterized by a sometimes rhythmically impressive movement restlessness of the head caused by myoclonia of the neck muscles involved in hyperkinesis.

1.1 Clinic

ST is a very rare disease. Frequency data are not found in the literature. GAUTHIER (1986) states in work on the pathophysiology and treatment of “idiopathic” ST, only gives the frequency of generalized dystonia for Canada at 4,2 per 1 million inhabitans, as he had no figures for ST itself. He presumably based this figure on new cases per year. ST is likely to occur far less frequently. In his own sick population men and women are represented in roughly equal numbers. Between 1979 and 1982, we diagnosed 115 patients, 59 women and 56 men with torticollis were examined and treated. Other authors found a slight preponderance of the male sex (HERZ and GLASER, 1949, SORENSEN and HAMBY, 1965, STEJSKAL, 1980). The average age at the onset of of the disease is given as 38 to 43 years (SORENSEN u. HAMBY, 1965, HAMBY and SCHIFFER, 1969, HERNESNIEMI and LAITINEN, 1977). The malposition of the head occurs more frequently lateralized to the left (ARSENI and SANDOR, 1959, STEJSKAL and TOMANEK, 1981). Familial occurrence has been reported (KRAYENBÜHL and YASARGIL, 1965, BOYSEN, 1979). GILBERT (1977) and FRIEDMANN and FAHN (1986) saw it in 9% of cases. This clinical picture is practically not observed in children (BOLTSHAUSER, 1976).

1.1.1 Symptomatology

The disease usually begins slowly and initially unspecific at first. A traumatic cause of the disease is generally not considered probable, although a cervical spine trauma is recalled in about one tenth of cases (BRÄUTIGAM, 1954, MARSDEN and SHEEHY, 1980). The patient initially complains of a stiff neck, sometimes associated with mild neck pain. In the initial phase the diagnosis of “cervical syndrome” is often made. However, the patient soon feels an involuntary pulling of the head to one side. As the disease progresses, an increasingly involuntary malposition of the head is noticed, often first by relatives. Finally, the full picture of ST develops with pronounced tonic or clonic tilting of the head in the form of a turn or tilt and visible hypertrophy of the sternocleidomastoid muscle (SCM muscle): Severe,

sometimes unbearable, cramp-like pain occurs, usually unilaterally in the neck region. The patient is then often no longer able to turn their head to the other side against the forced posture and the resulting resistance. Sometimes the head can no longer even be brought into a central position. Attempts to turn the head contralaterally are often associated with an increase in pain.

However, the symptoms are not always constant. Many patients experience relief when sitting with their head relief, as well as with the application of so-called tricks, e.g. the “Geste antagoniste”, already described by MEIGE and FEINDEL (1903). In this case, lightly placing 2 or 3 fingers on the side of the lower jaw opposite to the turn of the head reduces the pathological rotational impulses. According to WARTENBERG (1958), this attitude is called the “counterpressure phenomenon” in German. STEJSKAL (1980) analyzed this maneuver and observed that some patients also experience relief through “counterpressure”. However, a similar effect can also be achieved by raising both arms above head height. Patients often report that the head is completely calm when lying down and can be held straight; during sleep, the symptoms disappear completely, and only after getting up in the morning do the involuntary pulling and tension of the neck muscles set in again. The symptoms can be exacerbated by excitement, anxiety and other stressful situations, as well as by certain activities, e.g. walking or writing. The restlessness of the head then becomes even more pronounced. However, I have also been able to observe the opposite: In a 45-year-old female patient with a horizontal, tonic head turned 90 degrees to the right and not fixed head, the ST symptoms disappeared almost completely during rhythmic dancing or in an exposed situation, for example when giving a lecture.

The clinical picture of the disease is characterized by a more or less pronounced more incorrect posture of the head, which in in many cases is accompanied by an involuntary lack of movement restricted to the head and neck. The disease can therefore be classified clinically as a form of movement

disorders (hyperkinesia). This movement disorder is caused by mostly unilateral contractions of various neck muscles. Most commonly affected are the SCM and splenius muscles and the cranial parts of the trapezius muscles. Only rarely is involvement of the scalene group observed. In many cases, the deep neck muscles supplied by the upper 3 cervical roots are certainly also involved. Specifically, these are the semispinalis capitis and longissimus capitis muscles, as well as the rectus capitis posterior minor, rectus capitis posterior major, obliquus capitis superior and obliquus capitis inferior muscles around the short deep neck muscles. Involvement of the deep neck muscles can hardly be detected by palpation and is difficult to detect using EMG. However, analysis of the head deformity and the movement pattern suggest involvement of the deep neck muscles (Fig. 1).

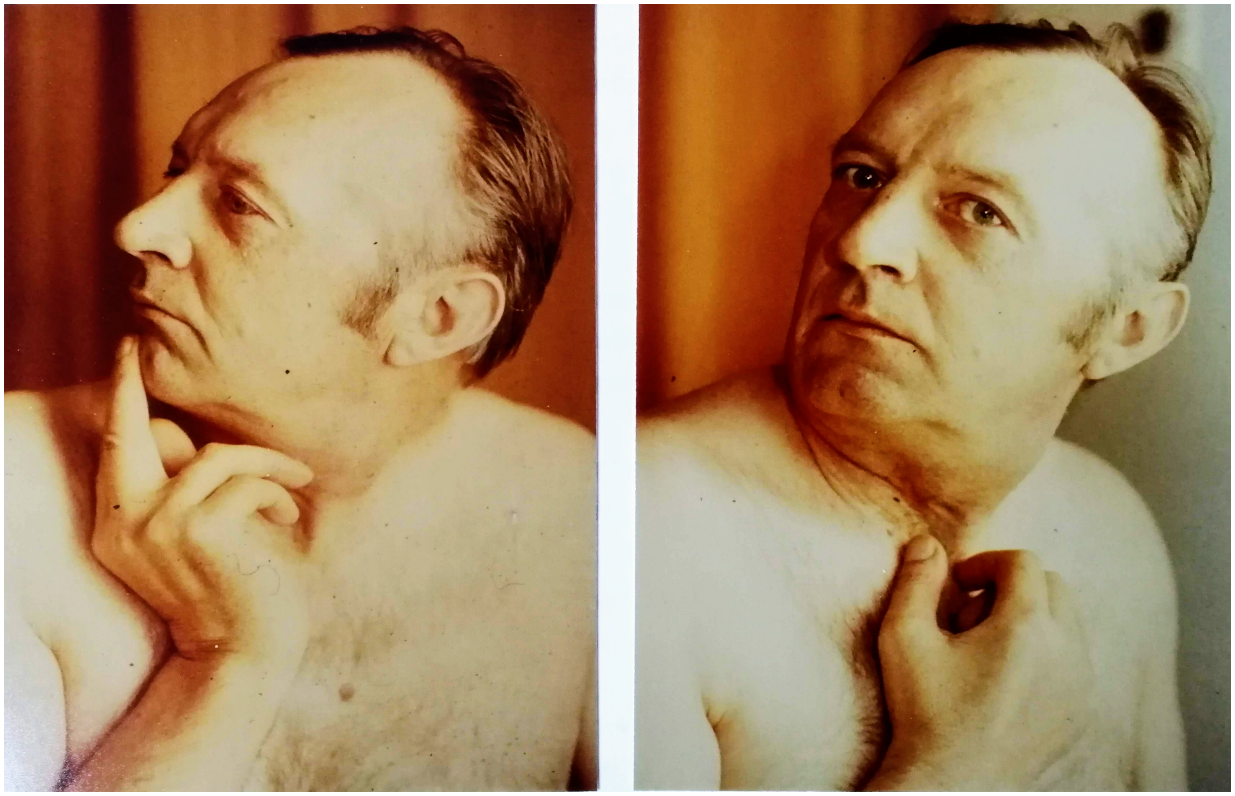

Fig. 1: Case 3, Horizontal ST to the left with “Geste antagoniste”.

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Depending on the type of head malposition, according to HASSLER and DIECMANN (1970), a distinction is made between the horizontal rotatory, anteflectory, retroflectory and combined horizontal-rotatory torticollis. The horizontal torticollis is defined as a turn of the head around the cranio-caudal axis, corresponding to the gaze turn in an upright primate. Rotation is defined as the inclination of the head around the fronto-occipital axis. Movements around the binaural axis (raising and lowering of the head) correspond to retroflexion and anteflexion. Horizontal and combined horizontal-rotatory torticollis are by far the most frequently observed. The purely rotatory manifestation of the symptoms is less common. Primary retrocollis is even rarer, while antecollis is practically non-existent in ST. It is most likely to be seen in postencephalitic or vascular forms of Parkinson's disease (HASSLER and DIECKMANN, 1970).
In addition to this primary symptom, in most cases a high shoulder develops on the side of the body to which the head turns or leans and on which the trapezius muscle is more involved. The contralateral SCM muscle, which is almost always predominantly involved in the symptoms, hypertrophies and is clearly prominent in relief. The clavicular tendon insertions are particularly tense, so that the involuntary muscle contractions can be felt particularly well here.

In the later course of the disease, which lasst for years, additionally secondary symptoms are added, such as cervical syndrome as a result of degenerative cervical spine changes (scoliosis) and finally also radicular disorders on the part of the cervical roots.

Due to the extraordinarily stigmatizing disease the torticollis patient soon comes under a strong suffering, which often leads to secondary psychological changes and severe social (family) stress, including suicidal tendencies (CURSCHMANN, 1907, FOERSTER, 1929, 1933, PATTERSON and LITTLE, 1943, HERZ and GLASER, 1949, PODIVINSKY, 1968, MATTHEWS et al., 1978).

Spontaneous regression of symptoms was observed more frequently in the first 3 years after the onset of the disease (BRÄUTIGAM, 1954, HERZ and GLASER, 1940, PATTERSON and LITTLE, 1943, JAYNE et al., 1984, FRIEDMANN and FAHN, 1986). The symptom-free intervals were usually only of limited duration, averaging about one year.

In summary, the clinical picture of pure ST is a hitherto etiologically unexplained disorder exclusively affecting limited to the muscles of the neck and throat, characterized by involuntary, mostly unilateral muscle contractions, which lead to an abnormal twisting or rotation, often accompanied by a clonic restlessness of movement, i.e. a skewed posture or rotation, i.e. tilting of the head.

1.1.2 Diagnostic equipment

The diagnosis of "ST" is made exclusively on the basis of clinical symptomatology. In "pure" ST, there are usually no other pathological findings apart from the EMG and secondary changes in the cervical spine (PODIVINSKI, 1968, THÜMLER, 1983).

1.1.2.1 X-ray diagnostics, computer tomography (CT)

Instrumental diagnostic procedures such as X-ray (SCHMIDT, 1978) and cranial CT are used to exclude a "symptomatic" torticollis, e.g. caused by a tumor, and the CT is normally unremarkable, without any indication of a possible cause of the condition (MURAYAMA et al., 1981, FRECKMANN et al., 1981, 1982, 1983, 1986, THÜMLER, 1983, VAN HOOF et al., 1987).

1.1.2.2 Cerebral angiography

Cerebral angiography sometimes can be used to detect vascular malformations and variants that could be involved in the pathogenesis of ST (SCOVILLE and BETTIS, 1979, FRECKMANN et al., 1981). Today we do not use angiography any longer, as it has been shown to have no significant influence on the choice of therapy.

1.1.2.3 Electromyography (EMG)

Most frequently the EMG is used to analyze the muscles involved in the symptoms. According to PODIVINSKY (1968), 3 types of torticollis can be distinguished 1. the tonic, 2. the clonic and 3. the mixed type. This typification was also adopted by LÜCKING (1980). VASILESCU and DIECKMANN (1975) distinguish between the SCM type, the splenius type and the trapezius type. This differentiation is primarily used to determine the target point for stereotactic interventions. Of great clinical importance is the fact that with ST, the agonists of both sides, e.g. the right SCM muscle and the left trapezius muscle are almost always involved in the symptoms at the same time, but not agonists and antagonists as in torsion dystonia (LÜCKING, 1980, LÜCKING et al., 1980, BERTRAND et al., 1978, 1982, HAGENAH et al., 1981, 1983).

IVANICHEV (1979) found changes in the EMG picture in depending on the duration of the disease. He attributed the lowering of the motor neuronal stimulation threshold to a secondary myopathosis of the muscles involved in the symptoms.

To analyze the muscles involved in ST and for the assessment of any tremor groups present, surface derivations were predominantly carried out (HERZ and HOEFER, 1949, PODIVINSKY, 1968, MEARES and LADER, 1970, VASILESCU and DIECKMANN, 1975, IVANICHEV, 1979, TOMANEK and STEJSKAL, 1979). In contrast, BERTRAND et al. (1987), HASSLER et al. (1981) and FASSHAUER (1980, 1983) used bipolar needle electrodes. The recording of individual motor units, as required for the assessment of potential size, width and shape, has so far only been published by HAGENAH et al. (1980, 1981, 1983).

1.1.2.4 Selective nerve/muscle blockade

Other methods for identifying the cervical muscles involved in ST are the selective blockade of the accessory nerve and the posterior branches of the 1st to 4th cervical nerves with a local anesthetic (RAMAMURTHY et al. local anesthetic

(RAMAMURTHY et al., 1978, BERTRAND et al, 1982, 1987) or the direct infiltration of these muscles with pancuronium (R) (CREMONESI and MURATA, 1986). This can also achieve temporary pain relief.

1.1.2.5 Electro-nystagmography

Electro-nystagmography may represent an enrichment of diagnostics in ST. TOMANEK and STEJSKAL (1979) and BRONSTEIN and RUDGE (1986) in the majority of their ST patients found asymmetrical nystagmus corresponding to the direction of rotation of the head after bilateral caloric vestibular irritation.

1.1.2.6 Positron emission tomography (PET)

Under the assumption that ST is a focal dystonia (adult onset dystonia), STOESSL et al. (1986) performed PET in 11 patients with pure ST and 5 patients with Dystonic torticollis (DT). They found no disturbance of the regional cerebral glucose metabolism. On the other hand, in their opinion, the findings were in favor of a bilateral disturbance of the normal relationships between the thalamus and the basal ganglia. The authors hypothesized that the cause of ST was an interruption of pallido-thalamic projection pathways with disruption in the neurotransmitting-system. However, the informative value of this method appears questionable, firstly because of the low resolution of a maximum of 14 mm and secondly because of the long examination time of 15 minutes per slice. Even CT, with considerably shorter scan times, is sometimes difficult to perform and assess due to the hyperkinesis of the head.

1.1.2.7 Evoked potentials (EP)

DISERTORI et al. (1982) were the first to report on the recording of auditory evoked brainstem potentials (AEP) in ST. In one case, they were able to demonstrate involvement of the brainstem, emphasizing mesencephalic structures. In contrast, NARAYAN et al. (1986) conducted a study on 10 ST patients in which they recorded visual and bilateral

somatosensory EPs from the median and peroneal nerves in addition to AEPs. They obtained normal responses in all recordings, both ipsi- and contralateral to the ST symptoms. According to the authors, these findings argue against a primary involvement of the visual, auditory and somatosensory pathways and control circuits in the genesis of idiopathic ST

1.1.2.8 Rheoencephalography

IVANICHEV and KHASANOVA (1979) investigated by rheoencephalography blood flow conditions in the vertebro-basilar tract in 22 ST patients. They found reduced blood flow in the area of the posterior fossa and in the neck muscles involved in ST. They postulated that the cause was compression of the neck vessels by the hypertonized muscles and by the vessels pressing against the cervical spine. When the head was in the middle position, however, the blood circulation improved significantly. No other studies dealing with this somewhat questionable method can be found in the literature, so that it is not possible to comment on the validity of this diagnostic procedure.

1.1.2.9 Pneumoencephalography (PEG)

The PEG no longer plays a role in diagnostics. It has been replaced by CT. However, the observations of HASSLER and DIECKMANN (1970), who frequently found ventricular asymmetries in ST patients, are noteworthy. They attributed these findings, which were not confirmed by other authors, to birth trauma or prenatal brain damage.

KASTE et al. (1981) found pneumoencephalographic in 10 of 13 ST patients fronto-parietally localized cortical atrophy. They concluded that fronto-parietal cortical lesions should be discussed for the etiology of ST.

VAN HOOF et al. (1987) found neither the ventricular asymetries described by HASSLER et al. DIECKMANN nor the cortical atrophy discussed by KASTE et al., so that these changes to be of no significance for the genesis of ST.

1.1.2.10 Electroencephalography (EEG)

All authors agree that the EEG has no diagnostic value. The brain wave pattern proves to be consistently unremarkable in pure ST.

1.1.2.11 Quantitative measurement of head turn and tremor

ANSARI and WEBSTER (1974) described a diagnostic method for measuring head rotation and any head tremor in ST. The analysis of the tremor recordings revealed two fundamentally different types of tremor, a regular tremor with 4 to 7 beats per second and a completely irregular tremor that varied greatly in frequency and amplitude. The relatively simple method is helpful in objectifying the clinical symptoms and is reproducible, so that the results of different forms of treatment can be compared with each other. VAN HOOF et al. (1987) measured the degree of head deviation in their torticollis patients in a similar way.

1.1.2.12 Magnetic resonance imaging (MRI)

At the end of this chapter I would like to mention MRI, which to my knowledge in the search for an organic substrate in the in the genesis of ST has not yet been used. Here urgently investigations are required, as MRI has already proven that it is particularly effective in regions of the brain, the brain stem and the medulla oblongata. However, the patient must lie absolutely still for a longer period of time, which in many cases would only be possible under anesthesia.

1.1.3 Other diagnostics

Normally, apart from ST symptoms, occasionally detected disorders of the vestibular apparatus and possibly secondary changes in the cervical spine, there are no other findings that deviate from the norm. There is no evidence in the literature to suggest a metabolic disorder. As a result, serum tests and other laboratory parameters do not allow any further conclusions to be drawn about the genesis of ST.

1.2 Differential diagnosis of ST

The differential diagnosis of ST has not always been given the importance, although the choice of therapy is closely related to it. This view is shared by many authors (CASSIERER, 1922, OLIVECRONA, 1931, KAPPIS, 1934, KRAYENBÜHL and YASARGIL, 1965, ARSENI and MARETSIS, 1971, COUCH, 1976, COLBASSANI and WOOD, 1986, BERTRAND and MOLINA-NEGRO, 1986, SHIMA et al., 1987).

In the differential diagnosis of typical ST, the muscular, ocular, and the possibly very rare psychogenic torticollis are important.

1.2.1 Muscular torticollis

The most common form of muscular torticollis is observed in young children. A unilateral shortening of the SCM muscle causes the head to tilt (rotate) to the diseased side and a head turn to the healthy side. The “congenital” torticollis is probably caused by perinatal damage. An intrauterine posture is also considered (VOLKMANN, 1885, LORENZ, 1895, MIKULICZ, 1895). SARNAT and MORRISSY (1981) suspected that the cause of this form of torticollis is ischemia of the sternal muscle head with a predisposed separate vascular supply of this of this part of the muscle. Sometimes a palpatory thickening in the lower third of the SCM muscle which usually resolves spontaneously over the course of weeks to months. In some children the SCM muscle must be surgically corrected (MIKULICZ, 1895, GARRE et al, 1930, RENTROP and STRASCHILL, 1981).

1.2.2 The osseous torticollis

The osseous torticollis is mainly found in congenital structural changes of the cervical spine (e.g. wedge vertebrae) or after traumatic after traumatic changes, such as cervical spine subluxation and cervical spine compression (RENTROP and STRASCHILL, 1981, MAXWELL, 1984). A rare cause of torticollis is the atlantoaxial instability of

inflammatory or traumatic origin (MAXWELL, 1984, ROOSEN, 1985, McCLELLAND et al., 1987). In contrast to ST, these forms of torticollis can be easily detected radiologically (DANDINE et al., 1980).

1.2.3 Torticollis in cervical syndrome

The malposition of the head that occurs in the context of acute cervical syndrome of the head, generally a fixed forced posture, is sometimes called “torticollis rheumatica” (GARRE et al., 1930, RENTROP and STRASCHILL, 1981, HÜLSE, 1983, MAXWELL, 1984).

1.2.4 The ocular torticollis

Ocular torticollis is usually the result of a congenital unilateral trochlear nerve palsy, which causes a compensatory compensatory tilting of the head. This prevents the perception of double vision (ELSCHNIG, 1929, WILSON, 1940, WALSH, 1957, BOLTSHAUSER, 1979).

1.2.5 Psychogenic (hysterical) torticollis

Psychogenic torticollis will be discussed in detail in the chapter of the ST pathogenesis hypotheses (see p. 20).

1.2.6 Rarer forms of torticollis

In addition to the forms of torticollis already mentioned, there are also a number of very rare clinical pictures, such as the torticollis in hiatal hernia, the pathogenesis of which is unknown, and paroxysmal torticollis in infants (RENTROP and STRASCHILL, 1981).

Infections in the cervical spine and the soft tissues of the neck can also lead to torticollis symptoms. VISUDHIPHAN et al. (1982) found 2 cases of retropharyngeal abscesses caused by streptococci. In 3 cases osteomyelitis in the cervical spine was responsible for the symptoms.

1.2.7 Symptomatic torticollis “spasmodicus”

Of fundamental importance in the differential diagnosis are “symptomatic” torticollis and the in context of torsion dystonia occurring “dystonic torticollis“.

1.2.7.1 Postencephalitic and postischemic torticollis

Numerous cases of torticollis can be found in the literature, some with the typical external appearance of ST, which are caused by various lesions of the central nervous system (CNS). The most well-known are postencephalitic forms of torticollis and those resulting from cerebral vascular events, e.g. infarcts of the basal ganglia (PATTERSON and LITTLE, 1943, GRINKER and BUCY, 1951, DENNY-BROWN, 1962, FAHN and BRIN, 1985).

1.2.7.2 Tumors as the cause of torticollis

AVMAN and ARASIL (1969) reported on a ST patient, in which the torticollis was caused by a colloid cyst of the 3rd ventricle. The symptoms regressed immediately after surgical removal of the cyst.

BOISEN (1979) reported three cases of ST caused by infratentorial tumors. One was a ependynoma tumor between the cerebellar tonsils and the medulla oblongata. The patient died as a result of tonsillar incarceration. Apart from the tumor, the neuropathological examination revealed necrosis in the area of the medulla oblongata with hemorrhage. The other two patients were members of the same family. Both the father and his son had a Lindau-tumor with large cerebellar cysts, one in the right cerebellar hemisphere and the other in the left cerebellar hemisphere.
In a paper on special clinical features of tumors at the level of the foramen magnum KRAYENBÜHL (1973) reported on a patient with a fixed head tilt to the right and simultaneous left spastic tetraparesis. The cause of this disorder was a left antero-lateral meningioma located at the level of the foramen magnum, which had led to a displacement of the spinal

cord. WALSH (1957) emphasized that torticollis is not uncommon in children with intracranial, especially infratentorial tumors.

JULOW (1983) reported 2 cases in which neurinomas of the spinal accessory nerve root (SANR) had led to torticollis symptoms.

KIWAK et al. (1983) reported on 3 children aged 9 months, 3 and 8 years of age, in whom a medullary tumor with syringomyelia and torticollis were found. VISUDHIPHAN et al. (1982) published 4 further cases in which the torticollis symptoms were caused by cervical medullary tumors.

1.2.7.3 Infectious torticollis

NENG et al. (1983) reported an epidemic occurrence of infectious torticollis in China. The initial symptoms of the disease, which resolved in all patients without any specific therapy, were fever, headache, cough, dizziness, vomiting and diarrhea. Although no pathogens could be detected, the authors suspected a viral disease with selective infestation of certain regions of the nervous system. In this context, FOX (1985) reported on a 3 1/2-year-old girl who fell ill with a flu-like infection and developed torticollis with head turning to the left. A viral infection was also suspected in this case. After the infection subsided, the torticollis regressed spontaneously.

MOORE et al. (1986) suspected that a defect in the immune system could play a role in these cases. In some ST patients they had observed a drop in of immunoregulatory lymphocytes.

1.2.7.4 Traumatic torticollis

Finally, I would like to cite a case of torticollis (SIMPSON, 1986) which, according to the author, may have been possibly caused by a glass splinter in the left side of the neck. As a teenager, the 32-year-old man had a stab wound on the left

side of his neck caused by a shard of glass. The right SCM muscle was hypertrophied. There was a horizontal torticollis symptomatology on the left. After surgical removal of the glass splinter, the symptoms had completely regressed.

1.2.7.5 Drug-induced torticollis

Drug-induced hyperkinesis (e.g. due to haloperidol or metoclopramide), which can mimic the picture of ST, disappear after discontinuation of the medication (THÜMLER, 1983).

1.2.8 Dystonic torticollis (DT)

The most difficult but also the most important differential diagnosis of ST is the differentiation from DT (HASSLER and DIECKMANN, 1979), with torticollis as the leading symptom of a central motor movement disorder, which is torsion dystonia. This torticollis can precede other movement disorders as a focal early symptom, which sometimes makes it impossible to differentiate it from ST. On closer examination, however, the movement disorder is usually found to extend beyond the neck muscles to the face and/or limbs. In these cases, it is a segmental torsion dystonia, such as the torticollis observed in conjunction with blepharospasm (MARSDEN, 1976, SCHENCK and SCHMIDT, 1978, JANATI, 1986), which is also referred to as “adult onset dystonia” (MARSDEN, 1976, 1985, SCHENK and SCHMIDT, 1978, FAHN and BRIN, 1985). Therefore, every torticollis patient must be examined for oro-mandibular dyskinesia and unilateral tremor of the arm or leg (Fig. 2). Sometimes the slight internal rotation of a foot may be the only sign of incipient torsion dystonia (MARSDEN and HARRISON, 1974, MARSDEN, 1976, 1985).

FOERSTER (1929, 1933) already emphasized that an athetosis could begin with the symptom “torticollis”. BRÄUTIGAM (1954) found in 20% of his cases with the leading symptom “torticollis” a later extension of the symptoms to torsion dystonia. He spoke of a transitory ST. BERTRAND (1987) observed 5 patients among 111 mostly “pure” ST cases in whom “adult onset dystonia” had developed from “classic” rotatory

torticollis within a few years. In this context, he pointed out, as did KRAYENBÜHL and YASARGIL (1965) and COUCH (1976), that the boundary between ST and focal dystonia is not always entirely clear.

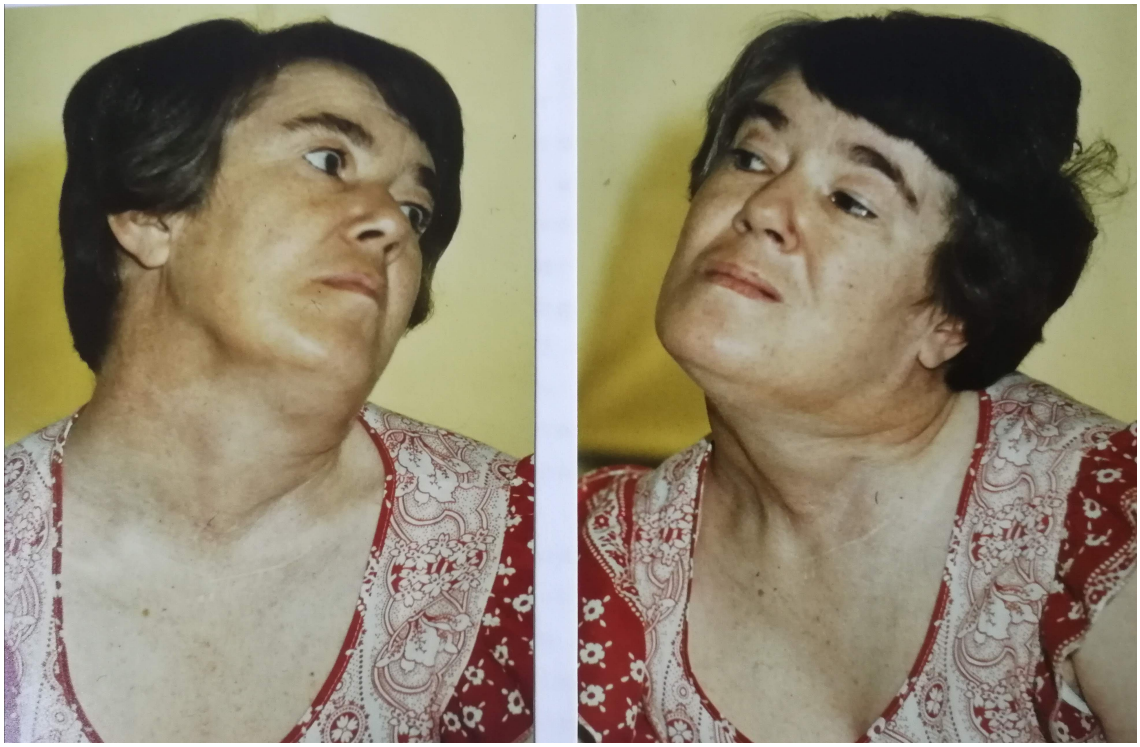

Fig. 2: Case 29: DT with combined torticollis symptoms (retrocollis with rotation).
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1.3 Aim of the paper

The aim of this paper is to describe ST and torticollis “dystonicus” (DT) as etiologically different clinical pictures, which accordingly also require a different therapeutic approach. While DT as a focal or segmental torsion dystonia remains classified as a central motor movement disorder, a lesion outside the CNS is postulated as the cause of ST. The hypothesis of a generally central genesis of ST is refuted. Instead, a peripheral neurogenic cause of “pure” ST is discussed.

The theory of an at least partially peripheral genesis of the of the disease is supported by electroneurophysiological, anatomical and surgical findings obtained from a patient population. The development of a new surgical approach for the treatment of ST based on a peripherally localized cause of the disease is described. The treatment results with this method are discussed and compared with the treatment and surgical results achieved to date.

The electroneurophysiological examinations were carried out by HAGENAH, who also assessed the EMG findings. This was based on, in addition to our own patient population, a stereotactically operated patient collective of MÜLLER from the years 1972 to 1987.

This work is divided into four parts. In the first part the various hypotheses on the pathogenesis of ST is discussed. In the second part the conservative and surgical treatment methods and their results are represent. The third part of the paper develops our own theory on the pathogenesis of ST and describes the new surgical method based on this and its results. The fourth part contains the casuistry as an appendix.

1.3.1 Problem definition

Psychiatrists, neurologists and neurosurgeons have always been faced with often almost unsolvable therapeutic problem.

The results of all the treatment methods that have been proposed to date are ultimately unsatisfactory because they are not causal. As a result, the methods of treatment that have been proposed to date are correspondingly diverse: Starting with psychotherapy, physical therapy measures and drug treatment attempts, they range all the way to a wide variety of largely destructive surgical methods, some of which target the brain. This shows the serious consequences for the patient if surgical treatment, e.g. stereotactic intervention is planned. Analogous to the different treatment methods, a wide variety of hypotheses have been developed regarding the aetiology of ST:

1. psychogenic triggering of ST,
2. the basal ganglia hypothesis,
3. the vestibular hypothesis and
4. the brainstem hypothesis (formatio reticularis)

PART I

2.0 The current hypotheses on the aetiology of ST

2.1 The psychogenic hypothesis

Towards the end of the last century, the term “torticollis mentalis” (BRISSAUD, 1895) was coined to differentiate it from ST. BRISSAUD was of the opinion that ST is only rarely a clinical picture with organic symptoms, but rather far more frequently a condition with a psychological (mental) basis (BRISSAUD and BAUER, 1909). This view was also held by KOLLARITS (1905, 1908), OPPENHEIM and JENDRASSIK (citation KOLLARITS, 1908), among others. KOLLARITS even thought that every ST, whether tonic or clonic, was a symptom of hysteria. To this day, the question of whether the disorder is of organic or psychological origin is sometimes controversial.

From a psychoanalytic point of view, MITSCHERLICH (1971, 1978), who has dealt with this problem in detail, considers ST to be a psychogenic disorder. It is a conversion syndrome (hysterical ST), but in some cases also obsessional neurotic or depressive forms of neurosis. In all ST patients, MITSCHERLICH found a body schema disorder with opposite-sex identification. Some patients showed that individual parts of the body, usually the neck, but also the entire upper body, had taken on the significance of the penis. MITSCHERLICH (1979) came to the conclusion that ST could be cured or at least improved by psychoanalysis and psychotherapy. PATERSON (1945), after examining 21 cases of ST, also came to the conclusion, as did ANDREWS and GILL (1982), that psychotherapy should be placed at the top of the list of possible treatment measures. Overall, however, the hypothesis of a fundamentally psychogenic triggering of the condition receives only little support in the literature.

Today, the view has generally prevailed that ST is predominantly a disease of organic origin, as has been demonstrated by several studies (HERZ and GLASER, 1949,

TIBBETS, 1971, COCKBURN, 1971, CHOPPY-JACOLIN et al., 1977, MATTHEWS et al., 1978). In most cases, the psychological changes observed in the patients were of a secondary nature (FOERSTER, 1920, 1929, PATTERSON and LITTLE, 1943, HERZ and GLASER, 1949, HASSLER, 1953, WYCIS and MOORE, 1954, CHOPPY-JACOLIN et al., 1977, van HOOF et al., 1987).

MEARES (1970, 1971, 1973) also accepted the organic genesis of the disease for the majority of ST patients. In a small number of cases, however, he found with the help of psychiatric and psychoanalytical examinations, indications of a psychogenic cause of the disease. In these cases, surgical treatment could not be helpful.

van WAVEREN (1982) demanded that the ST patient, before choosing the therapeutic approach, both neurological as well as psychiatric examination in order to be able to avoid surgery in psychogenic torticollis cases. In severely depressed patients, the indication for surgical intervention should be made very cautiously, as the success of treatment is very much in question in these patients (WITZMANN et al., 1984).

In summary, however, it can be stated in agreement with most authors that, with a few possible exceptions, an organic cause should generally be assumed in ST (CURSCHMANN, 1907, FOERSTER, 1929, HERZ and GLASER, 1949, WYCIS and MOORE, 1954, PODIVINSKY, 1969, MATTHEWS et al., 1978, BERTRRAND et al., 1978, MARSDEN, 1985 and many others).

2.2 The vestibular (labyrinthine) hypothesis

In the context of the scientific dispute as to whether the ST, as claimed by KOLLARITS (1905), as an exclusively “hysterogenic product”, CURSCHMANN (1907) came to the conclusion that in the case of torticollis an organic cause has to be investigated. He justified this postulate with the observation that a unilateral labyrinthine disease could lead to the typical picture of ST.

At the age of 19, a 37-year-old man had developed right-sided chronic ear disease at the age of 19, which led to deafness. 10 years later “seizures” occurred, which began with severe right-sided ringing in the ears and then led to nausea and severe dizziness. 5 years later, a typical ST developed with a head tilt to the left. Except for the function of the accessory and the statoacusticus nerves, the neurological findings were normal. The newly introduced quinine treatment had led to complete freedom from symptoms, which lasted for 1 1/2 years.

In the second case, a 59-year-old woman, the symptoms were similar: She suffered from tinnitus and a horizontal rotary vertigo that occurred intermittently. Gradually, a clonic torticollis developed with a head turn to the left. Otherwise unremarkable neurological findings. The otologic examination revealed a mild hypacusis on both sides without major labyrinth disorders. However, there was an increasing horizontal nystagmus to the left with the rotatory vertigo. Here, too, there was a temporary improvement in symptoms after quinine treatment.

The third case, a 28-year-old woman, also developed severe attacks of horizontal rotary vertigo with tinnitus and increasing hearing loss. At the same time, a clonic torticollis with turning of the head to the right and slight retroflexion occurred. Otologically, a slight hypacusis was found on both sides with chronic tubal catarrh. Otherwise normal neurological findings. Here, too, the quinine treatment had improved the symptoms.

CURSCHMANN (1907) noted “that the spastic nature of torticollis, in the in contrast to an ordinary corrective posture, is shown by the fact that the torticollis does not disappear immediately after removal of the provoking stimulus, but that the posture and tension of the spastic muscles only improve very gradually over the course of a few weeks”. In conclusion, CURSCHMANN states that his observation prove “that labyrinthine diseases can lead to a ST due to the dizziness they produce in certain directions, in which mainly

the muscles supplied by the accessory nerve are involved”. While CURSCHMANN is of the opinion that ST is a disease of organic origin was able to assert itself, his hypothesis of a labyrinthine genesis of ST found little support. Only HYNDMANN (1939) and WILSON (1940) shared this view. They considered a primary involvement of the vestibular system in the pathogenesis of ST, as they also found pathological labyrinth functions in some of their ST patients.

TOMANEK and STEJSKAL (1979) conducted electronystagmographic investigation on 40 ST patients, also with the question of whether there is a connection between this disease and the vestibular apparatus. Although the significantly high nystagmus values were detected in the caloric test, the vestibular asymmetries, although correlating with the direction of the ST, did not reach pathological values, so that a primary vestibular involvement must be regarded as unlikely.

Vestibular involvement was initially supported by studies by BRONSTEIN and RUDGE (1986) on 35 ST patients. With otherwise unremarkable neurological and otological findings, they found electronystagmographically in 70% of the cases vestibular nystagmus that was opposite to the direction of the ST. With the possibility of optical fixation, nystagmus was less frequent and less pronounced. To the same extent, the calorically tested labyrinth function was pathological. The authors concluded that a primary involvement of the vestibular system could not be excluded in ST. A disturbance of central, stimulus-processing mechanisms, which are responsible for the orientation of the head and eyes in space, had to be discussed. Only recently BRONSTEIN et al. (1987) relativized this view again: There must be two different forms of ST, a torticollis after vestibular damage and a form of ST without primary vestibular involvement.

MATTHEWS et al. (1978), on the other hand, apart from exeptions considered a vestibular or labyrinthine genesis of ST as completely improbable, since no pathological labyrinthine functions were detectable in their patients

despite detailed examination, so that this hypothesis was apart from individual cases, which, however, must be attributed to the symptomatic forms of torticollis, this hypothesis must be regarded as unproven and hardly probable.

It therefore ultimately remains unclear whether the symptoms electronystagmographically proven central disturbances of the vestibular system are of a primary nature and are therefore cause of the ST or whether they are the result of asymmetrical afferent impulses from head joints (DIAMOND et al., 1987).

2.3 Basal ganglia hypothesis

As early as 1920, FOERSTER expressed the hypothesis that ST was a disease of the basal ganglia. In athetotic clinical pictures, where a lesion in the area of the neostriatum (corpus striatum, caudate nucleus, putamen), he often observed torticollis. Based on this, he assumed a localized athetosis in ST, which remained limited to the neck muscles. He suspected focal lesions in the part of the neostriatum in which the neck muscles are represented (FOERSTER, 1929, 1930, 1933). FOERSTER (1933) emphasized his view with a dissection case: It was a 23-year-old man who, however, did not suffer from a typical ST, but from an extreme retrocollis due to bilateral, symmetrical spasms of the neck muscles. The anterior and posterior roots of the 1st, 2nd and 3rd cervical nerves on both sides were severed intradurally. Postoperatively, an extensive epidural hematoma developed in the surgical area, as a result of which the patient died. The pathological anatomical examination of the brain revealed several small lesion foci in the area of the putamen, which he held responsible for the clinical picture: “Torticollis can be the only symptom of a striated disease. If the brake (corpus striatum) applied to the thalamopallidal reflex arc is removed, the excitatory current flowing to the thalamus pours uninhibitedly into the periphery via the pallidum, resulting in continuous involuntary muscle innervations which appear as an athetotic play of movements on the plan.” FOERSTER (1929) continues a little later: “The striated

torticollis can be considerably intensified by all possible intercurrent sensory or motor stimuli as well as by affects. In particular, irritative processes in the cervical region or in the vestibular apparatus seem to have a spasm-increasing and spasm-sustaining effect”.

This hypothesis, in which the ST is regarded as a “focal” or “localized” torsion dystonia, to the neck muscels restricted, has been adopted by most authors to date. Focal dystonia, in the in the context of extrapyramidal motor movement disorders, include, in addition to ST, blepharospasm, oromandibular dyskinesia and dyskinesia and writer's cramp (MARSDEN, 1974, 1976, 1985, LANCET (EDITORIAL) 1978).

The most important representatives of this hypothesis are listed here: COOPER, 1965, HASSLER and DIECKMANN, 1970, 1971, COUCH, 1976, MARSDEN, 1976, 1985, GOLDHAHN and GOLDHAHN, 1977, BERTRAND et al, 1978, MOLINA-NEGRO, 1979, POSER et al, 1979, ANDREW, 1981, SCHMIDT and POTTHOFF, 1981, OLANOW, 1981, KOREIN et al, 1981, KOREIN, 1981, THÜMLER, 1983, FAHN, 1985, and GILLMAN and SANDYK, 1985. These are almost exclusively researchers who use their therapy, stereotactic surgical procedures or forms of drug treatment targeting the neurotransmitter system of the basal ganglia region, are based on this hypothesis.

Almost all efforts to find a neuropathological substrate for the for the etiology of ST have therefore been based on the the idea that this must be sought in the basal ganglia region. Therefore I would like to summarize the published neurophathological findings and add a further case of my own.

2.3.1 Pathological anatomy

Overall, only a few dissection findings have been published. The main reason for this is that ST “per se” does not lead to death. These cases are therefore mostly patients who died as a result of postoperative complications. Furthermore, examination of the findings reveals that the clinical symptoms were rarely limited to the neck muscles. Here, too,

it can be seen that cases were often included under the diagnosis of “ST” which should have been assigned to the diagnosis of “DT” or “torsion dystonia” if strict criteria had been applied.

Case 1 (KOLLARITS, 1908): The author reported on a man who, after a typhoid infection, 5 years before death, first became ill with cramps in the feet, then in the trunk and finally in the neck combined with head tremors and grimacing. Initially, the symptoms could be favorably influenced by the “Geste antagoniste” and conservative treatment measures. Then there was an increasing worsening. A “sham operation” was unsuccessful. Finally, the patient died of exhaustion at the age of 44. The neuropathological examination revealed no abnormalities apart from a degeneration of the posterior cords in the area of the medulla oblongata. KOLLARITS himself doubted that the torticollis was related to the described spinal cord degeneration.

Case 2 (CASSIERER, 1922): The case concerned a man who died at the age of 29 after myotomy of the deep muscles of the neck. Already at the age of 7 years gait disturbance with internal rotation of the left foot. In the course it gradually developed into the full picture of a torsion dystonia with pronounced retrocollis. Myotomies and nerve transections of the neck and extremities were performed several times without success. In addition to brain swelling, the autopsy revealed “undoubted signs of decay in the ganglion cells, neuronophagia, amoeboid glia and moderate capillary fibrosis”. These changes were most pronounced in the corpus striatum, but to a lesser extent also in the thalamus and in the cerebral cortex. Although a psychogenic cause of the disease was clinically suspected in the patient, the pathological-anatomical examination certainly revealed an organic substrate. CASSIERER writes: “To establish a closer relationship between the anatomical changes found and the clinical picture seems to me to be completely impossible for the time being.”

Case 3 (WIMMER, 1929): The case concerned a 16-year-old girl

who had developed normally up to the age of just 5 years. Then epileptiform, markedly clonic convulsions occurred for the first time. Finally, the typical picture of a pronounced torsion dystonia with strong involvement of the head in the symptoms developed. The cause of death was not reported. Neuropathologic examination revealed striking microgyria of both frontal brain poles, dysplasia of the right cerebral hemisphere, with a large cyst extending from the right frontal pole to the lower portions of the right parietal lobe, complete absence of the anterior portion of the right caudate nucleus, and more posteriorly pronounced atrophy of the right putamen and pallidum. Microscopic examination revealed lesions in the right neostriatum associated with glial proliferation.

Case 4 (FOERSTER, 1933): This brain dissection finding has already been described above.

Case 5 (GRINKER and WALKER, 1933): This was a 25-year-old female patient suffering from a ST with involuntary turning of the head to the right. The duration of the history was 5 years. The history was otherwise unremarkable. The clinical symptoms were limited to the head and neck. During the operation, the upper posterior cervical roots and the third anterior cervical root on the right were severed. Due to severe venous bleeding, the procedure had to be aborted. 8 days later the wound was reopened to complete the rhizotomy. This resulted in a delirious drop in blood pressure. The patient died a few hours after the operation. The neuropathological examination revealed hyperplasia of the meninges and perivascular round cell infiltrates throughout the brain as evidence of chronic encephalitis. The ganglion cells of the cortex and basal ganglia, especially in the caudate and putamen areas, showed signs of chronic degeneration with regressive glial changes. There was no evidence of a specific lesion that could have been responsible for the ST.

Case 6 (ALPERS and DRAYER, 1937): A 90-year-old man suffered

since the age of 43 from a compulsive head to the right, oro-mandibular dyskinesia and choreiform hyperkinesis of the upper extremities. Death was due to old age. Brain dissection revealed fibrotic thickening of the arachnoid membrane, atrophy of the neostriatum with marked changes in the large ganglion cells in the caudate and putamen areas. The changes in the pallidum were less pronounced.

Case 7 (SOLCHER, 1957): The patient was a 81-year-old woman who had been suffering from the consequences of of prenatal carbon monoxide poisoning. In addition to a pronounced Parkinson's syndrome, there was a beating retrocollis. With severe cachexia, death occurred as a result of cardiovascular failure. The brain section revealed a status marmoratus in the entire neostriatum. The The most conspicuous changes were seen in the pallidum.

Case 8 (TARLOV, 1970): In a 65-year-old woman developed a ST with turning and tilting of the head to the left 6 years before death. Involuntary movements of the head occurred mainly when standing and walking. The psychiatric examination revealed no abnormalities. The otologic examination suggested a slight hearing loss on the left, while the vestibular examination revealed no pathological findings. The woman died as a result of bronchopneumonia. Brain dissection showed no pathologic findings other than mild thickening and mononuclear infiltration of the meninges.

Case 9 (TARLOV, 1970): In the same paper TARLOV reported a second ST case, which was dissected by GREENFIELD in 1936. No special features were found here. TARLOV pointed out in this context that animal models for the production of abnormal head postures may not correlate with the pathological anatomical substrate in humans.

Case 10 (LOZANO-SAAVEDRA, 1979): Of 23 ST patients undergoing stereotactic surgery, one patient died 21 days after the procedure as a result of pulmonary embolism. A second patient committed suicide after further deterioration of his symptoms with combined rotatory and retrocollis. The neuropathological

examination revealed no evidence of striatal damage in either case. However, meningeal thickening was conspicuous. For the first time it was explicitly emphasized here that an examination of the deeper brain stem sections was also carried out. However, here too, no pathological changes were found that would have allowed conclusions to be drawn about the cause of the disease.

Case 11 (LOZANO-SAAVEDRA, 1979): See case 10.

Case 12 (see case history, case 37): This was a 43-year-old male patient who, 4 years before his death, developed a slowly progressive combined horizontal-rotatory ST symptoms with turning and tilting of the head to the right. Two years after the onset of symptoms, a left subthalamotomy was performed. Postoperatively, the torticollis symptoms were essentially unchanged. However, as a side effect of the operation, a discrete, arm-emphasized hemiparesis on the right with a disturbance of fine motor skills was added. In addition, a speech disorder and a reactive depressive mood developed. 8 months after the stereotactic procedure, we performed the BMLA.

The postoperative course initially was complicated by a massive abacterial meningitis. At the final examination, however, a slight improvement in the torticollis symptoms was noted. In the course of the following year, with already existing spondylotic changes in the cervical spine, a C4 root irritation syndrome on the left developed with severe pain radiating into the arm. The ST symptoms appeared improved, but by no means regressed. Therefore, 10 months after exposure of the craniocervical junction, the surgical site was revised.

Postoperatively, the patient initially recovered slowly, but could finally be fully mobilized. Two weeks after the operation, an internal hydrocephalus developed internus developed, which was drained. However, the patient's condition continued to deteriorate with signs of increasing respiratory insufficiency. Mechanical ventilation became

necessary. Finally, pneumonia led to the development of pulmonary fibrosis with a corresponding diffusion disorder. Death occurred three months after the operation.

The neuropathological examination revealed no pathological findings on the brain, particularly in the area of the basal ganglia, apart from the stereotactically caused subthalamic lesion (Forel's field) on the left and a clear ventricular enlargement. In the area of the medulla oblongata pronounced dorsal convolutions were found. As far as the postoperative and ischemic changes, no pathological changes were recognizable in this area that would have allowed a conclusion as to the cause of the disease.

2.3.1.1 Summary of the autopsy findings

The compilation of these few neuropathological findings alone shows that the view that ST is caused by lesions in the area of the neostriatum is on a very shaky ground. A critical appraisal of the findings shows that damage to the striatum was only present in those cases in which the torticollis was a partial symptom of athetosis or torsion dystonia (cases 1, 2, 3, 4, 6, 7). In the “pure” ST cases, the hyperkinesis was limited to the neck muscles, neuropathological findings have not been described (cases 5, 8, 10, 11, 12) (COLBASSANI and WOOD, 1987).

2.3.2 Torticollis in animal experiments

In addition to efforts to find a pathological anatomical substrate, in the hope of obtaining information on the genesis of ST, various animal studies were also carried out to produce torticollis-like symptoms. Only stereotactic procedures were used for this, with target points in the basal ganglia, the mesencephalon, but also the vestibular nuclei and the formatio reticularis. “Stereotactic” means spatially targeted and describes a surgical technique in which a probe is inserted into a circumscribed area of the central nervous system via a high-frontal borehole using a targeting device attached to the outside of the head. After

electrical stimulation of the deep-lying brain structures, in most cases the basal ganglia, the desired target point is reached. At the target point, a circumscribed coagulation and destruction of the brain tissue is then induced by heating the tip of the probe. This leads to an interruption of defined pathways in the extrapyramidal system. As these experiments are predominantly based on the basal ganglia hypothesis, I would also like to discuss them here.
As early as 1925, BERNIS and SPIEGEL pointed out the importance of the basal ganglia and the formatio reticularis for the static innervation and for the regulation of skeletal muscle tone. SPIEGEL (1927) wrote: “The essential nature of the static innervation of vertebrates, which is essentially maintained by reflex permanent excitation, is brought about by impulses from a wide variety of sources. Among these are excitations from the musculature of the limbs, from the muscles of the neck and from the labyrinth. In addition excitations from the body surface and from the retina are of secondary importance. The permanent excitations entering the spinal cord via the posterior roots partly maintain intraspinal reflexes, partly they draw to the tonus centers located in the formatio reticularis. Here in the rhomboid brain, therefore, the labyrinthine nuclei on the one hand and the reticular formation on the other represent the main centers of the tone-regulating supraspinal reflexes.”
In cats, HESS (1941, 1956) was able to trigger rotational movements of the head around the longitudinal axis of the body on the one hand and raising or lowering of the head on the other by stimulating and deactivating certain areas in the basal ganglia.

Inspired by these studies, HASSLER (1956) in cats carried out irritations on a direct vestibulo-thalamic pathway in the area of the pons, the midbrain and the thalamus. This generated ipsiversive movements in the animals of the head, but also simultaneously of the entire body. After switching off these structures, identical but contraversive turning movements. HASSLER therefore made the following observations: While turning movements in lower mammals were performed with the whole body, in higher mammals they were mainly performed

with the head. In primates, instead of the body or head turn can even be replaced by a turn of the “gaze apparatus”. In the ascending mammalian series, the head as the carrier of the visual apparatus gains increasing freedom of movement in relation to the trunk. In humans, the eyes can even without changing the position of the head in a wide angle. He concluded that the horizontal gaze movement is only a special case of head-turning movements. The formatio reticularis is the selective organ upstream of all effectors for all kinds of turning movements, including gaze turns. It integrates the various systems that are necessary for movements in the horizontal plane with the proprioreceptive afferents, especially from the cervical region. HASSLER describes these systems as the “central apparatus of turning movements”.

FOLTZ et al. (1959) relativized these investigations and the view that the conditions in the cat could hardly be were hardly transferable to humans. They therefore carried out experiments on monkeys (Macaca mulatta). In 6 animals they applied stereotactically more extensive electrolytic lesions in the medial parts of the mesencephalic part of the formatio reticularis at the level of the brachium conjunctivum. In one animal only a stimulation electrode was implanted at the same site. In all animals a persistent, predominantly clonic torticollis contralaterally, during stimulation ipsilateral to the lesion site could be generated. According to the authors, the torticollis symptoms were similar to the clinical picture of ST in all cases and was relatively constant from animal to animal. However, in most animals FOLTZ et al. concluded that the ST was caused by a disorder in the medial reticular formation at the level of the mesencephalic tegmentum, with interruption of certain pathways to neurons responsible for controlling head posture. Analogous to the treatment of other hyperkinesis they hypothesized that it must be possible to influence the ST better stereotactically than with peripheral interventions.

Later, HASSLER and DIECKMANN (1968) obtained the following results in cats after stimulation of the putamen ipsilateral turning movements. The results of putamen stimulation were in

favor of an ipsilateral inhibition of the pallidum, so that the impulses of the contralateral pallidum dominated. Stimulation of the pallidum therefore led to a contraversive turn of the body.

MORI et al. (1975) also carried out experiments on cats. They stereotactically destroyed areas in the paramedian mesencephalic tegmentum. About two thirds of the animals showed torticollis-like head postures. In addition to the histological analysis of the basal ganglia, the concentration of noradrenaline and dopamine were measured separately in the basal ganglia. MORI et al. found a strong drop in serotonin on the damaged side. From this they drew conclusions for the clinic: they reported that the medication with amantadine (Symmetrel (R)) in 7 out of 8 ST cases in which stereotactic treatment was unsuccessful led to an improvement in symptoms.

Later, MORI et al. (1979, 1985) produced in cats a chemical destruction of the mesencephalic tegmentum in cats. In the assumption that the decrease of serotonin in the neurotransmitter system of the basal ganglia plays an essential role in the genesis of ST, stereotactic neurotoxic reagents were injected into the brain tissue. However, this did not produce any abnormal movements. The authors came to the conclusion that lesions in non-dopaminergic structures had to be held responsible for the manifestation of torticollis symptoms.

In contrast, CROSSMANN and SAMBROOK (1978) were able to demonstrate in monkeys, after unilateral injection of 6-hydroxydopamine into the lateral hypothalamus with interruption of the ascending nigrostriatal, dopaminergic pathway, a provoke of torticollis symptoms. They concluded that there is a connection between disturbed dopamine metabolism and ST.

Similar experiments were carried out on cats by MALOUIN et BEDARD (1983). In this context, the authors pointed out the importance of the formatio reticularis and its ascending

pathways to the caudate for the control and regulation of head symmetry.

The attempt by GOPALAKRISHNAKONE (1985) to link the skewed posture of neck and head observed in Peking ducks with the human ST seems questionabel. The microscopic examination of the neck muscles revealed showed differently pronounced signs of muscle degeneration with signs of inflammation.

2.3.2.1 Summary of the findings from animal experiments

An experimentally induced head malposition in lower mammals can hardly be transferred to humans, because in phylogenesis the positioning function of the midbrain apparatus can be inhibited more and more arbitrarily in higher animals, an ability that is much more pronounced in humans (SCHALTENBRAND, 1925, SPIEGEL, 1927, LOZANO-SAAVEDRA, 1979). TARLOV (1970) pointed out that animal models for the production of abnormal head postures may not correlate with the pathological anatomical substrate in humans. Stereotactic stimulation and deactivation are much more likely to be a model for postischemic or postencephalitic athetosis. In healthy animals, artificially induced unilateral lesions in the extrapyramidal motor system, at any site, lead to a unilateral change in muscle tone. However, similar effects can also be achieved by labyrinthectomy (McCOUCH et al., 1951) or by unilateral transection of the posterior root of the 1st cervical nerve (HÜLSE, 1983). Therefore, only experiments on primates in which the control of head motor function, including the corresponding highly complex control circuits, is developed in a similar way to humans would appear to make sense. However, such experiments have not yet been carried out.

2.3.3 Other findings

HASSLER and DIECKMANN (1970) by PEG found ventricular asymmetries in ST patients, which they interpreted as an expression of basal ganglia damage, while KASTE et al. (1981) found unilateral cerebral cortical atrophy, which they

considered a possible indicator of a central motor cause of ST. These findings could not be confirmed by CT. The cranial CT normally is completely unremarkable, especially a lesion in the area of the basal ganglia that could be the cause of ST has not yet been described (THÜMLER, 1983, FRECKMANN et al., 1981, 1982, 1983, 1986, VAN HOOF et al., 1987).

Against the involvement of the basal ganglia also speaks the analysis of visual, auditory and somatosensory evoked potentials on 10 ST patients by NARAYAN et al. (1986), which revealed completely normal findings.

The reported results of a PET study by STOESSL et al. (1986), according to which ST is associated with a disorder in the neurotransmitter system of the basal ganglia must be considered questionable. The resolution of this examination technique is too low and the influence of disturbances in a neurophysiological control circuit on the metabolism is too little known to draw conclusions about a metabolic disorder in the basal ganglia.

It should be noted that neither the above-mentioned findings and animal experiments mentioned above, nor all other investigations examinations carried out to date on the ST provide evidence for the the correctness of the basal ganglia hypothesis.

2.4 The formatio reticularis hypothesis

As already mentioned, BERNIS and SPIEGEL (1925) established that for the static innervation and maintenance of muscle tone static innervation, apart from the vestibular nuclei, the formatio reticularis is responsible. Here ascending proprioceptive stimuli are converted into efferent impulses together with afferents from the vestibular system.

Based on the observation that in diseases in the posterior fossa, especially in connection with tumors in the cerebellum, sometimes a tilt of the towards the affected side apears, KEMBERLING, et al. (1952), with the collaboration of

SPIEGEL, were the first to produce an experimental “torticollis” in non-decerebrated cats. Stereotactically, they applied unilateral lesions in the vestibular nuclei and in the neighboring reticular formation. This resulted in a persistent “torticollis-like”, strong inclination and rotation of the head to the operated side. They concluded from this that the reticular formation is not only of great importance for the static innervation of the skeletal musculature, but above all for the control of head posture.

The position of the head therefore is determined by a large number of rhombencephalic reflexes. Proprioceptive afferents from the neck muscles, the cervical spine joints, especially the head joints, vestibular afferents, optical and acoustic sensory stimuli are processed and coordinated in the formatio reticularis in a complex manner (LOZANO-SAAVEDRA, 1979, NIEUWENHUYS et al., 1980) (see Fig. 3).

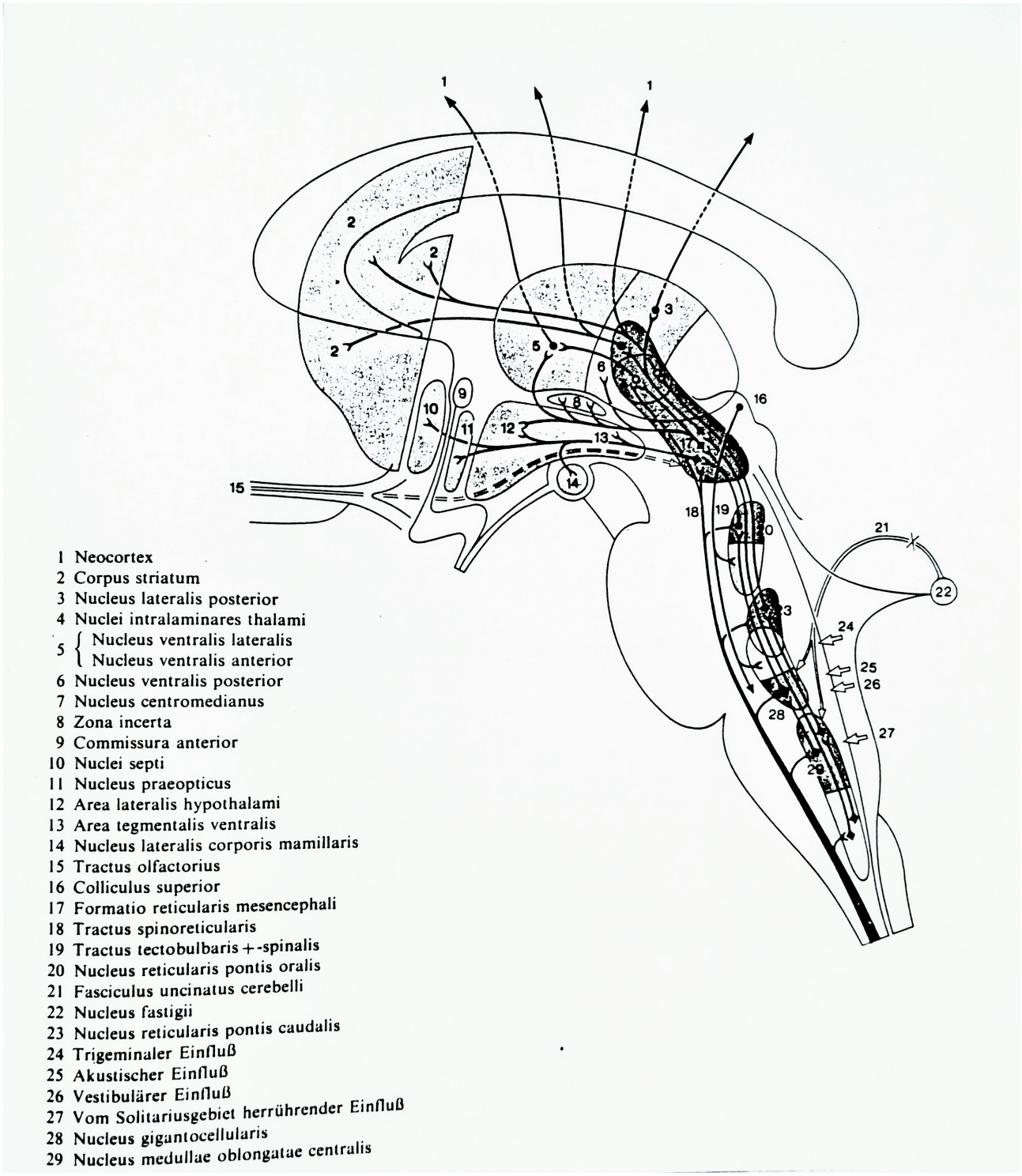


Fig. 3: Ascending fiber systems of the formatio reticularis and vestibular afferents (From NIEUWENHUIS et al. (1980) The human central nervous system).

Only more recently, based on these neurophysiological principles, the formatio reticularis hypothesis was developed, which is closely linked to the idea of a vestibular genesis of the ST.

SVIEN and CODY 1969 published a case similar to the CURSCHMANN cases: A 50-year-old woman had 10 years earlier an episode of vertigo lasting about 1 month. The same event occurred again 9 years later. 6 months later, a typical ST developed with head turned to the left. The right SCM muscle was hypertrophied. The otologic examination revealed a nystagmus to the left. The remaining neurological findings were normal. After bilateral iontophoresis of the middle ear with a mixture of hyaluronidase and tolazoin, the symptoms regressed. The same treatment also produced excellent results in 6 other cases.

Apparently unaware of the work of CURSCHMANN (1906), SVIEN and CODY came to the following hypothesis: The fact that the vestibular end organs have both afferent and efferent connections with the vestibular nucleus, that the vestibular nucleus receives inhibitory impulses from the formatio reticularis and that the vestibular system is strongly influenced by spinal reflexes, any change in the normal balance of this system must lead to activity of the neck muscles. As a result, small lesions in the area of the formatio reticularis can lead to an interruption of the inhibitory influence on the same-sided vestibular nucleus and that the balance of the cervical reflexes can be restored by reducing the impulses from the utriculus to Deiter's nucleus. The authors noted, “If ST is viewed not as a basal ganglia disorder (a view with little support) but as a kind of counterpart to ocular nystagmus, which manifests in the neck muscles, then attention is drawn to the labyrinth, which may be involved in the development of this disorder.”
The formatio reticularis hypothesis found further support by SIROKY et al. (1978, 1980), who reported 18 good treatment results after iontophoresis in 22 ST patients. Two patients suffered a recurrence, in 2 other patients the treatment had no effect.

LOZANO-SAAVEDRA (1979) presented in detail the hypothesis of a triggering of the ST by dysfunction of rhombencephalic neuron groups, in particular undifferentiated neurons of the reticular formation close to the binder arms, which could be assumed to serve as a substrate for the formation of complicated reflexes: “The reticular formation coordinates the contraction of the agonists and the inhibition of the antagonists of the neck muscles”. He concluded “that the ST is based on very small, possibly only quantitatively detectable changes in the area of the reticular formation”. However, careful pathological examinations of the deep brain stem in two dissection cases revealed no changes that could support this hypothesis.

The following considerations also argue against a cause of the reticular formation: Iontophoresis of the middle ears eliminates the vestibular end organs. This is therefore a change in the peripheral input to the vestibular nuclei. However, the sense of balance is by no means only influenced by the semicircular canals. Rather, it involves serveral “redundant” organs for maintaining balance, which are linked in a complex way in the formatio reticularis of the lower brain stem. In addition to the afferents from the vestibular organs, these include afferents from the visual system, from the eye muscle nuclei, from the head joints, the upper cervical spine joints and from the tension tension receptors of the neck muscles, especially the SCM and trapezius muscles. To a lesser extent, the proprioceptors of the rest of the body are also involved (MAGNUS and DE KLEIJN, 1912, BERNIS and SPIEGEL, 1925, SPIEGEL, 1927, DUENSING and SCHÄFER 1960).

The peculiarity of this complex system can also be seen in the difference between the SCM muscle and the upper trapezius to the rest of the skeletal musculature. These predominantly branchiogenic muscles (McKENZIE, 1955) are particularly dense with muscle spindle afferents (ABRAHAMS et al., 1975, FITZGERALD et al., 1982). In comparison to lower mammals, where the SCM muscle is purely branchiogenic, the different structure of the SCM muscle and the upper trapezius in humans

is particularly clear (McKENZIE, 1955). As with the external eye muscles, the laryngeal muscles and the diaphragm, these are parallel-fibered lifting muscles. It can therefore be said that they morphologically and functionally a middle position between the the outer eye muscles and the other feathered skeletal musculature.

ZANGEMEISTER et al. (1980, 1981) in this respect found that the SCM muscle, the upper part of the trapezius, but also the splenius muscle, are innervated before the outer eye muscles during a reflexive gaze that goes beyond the degree of freedom of the bulbi. These muscles therefore can be functionally described as secondary eye muscles which, controlled via the reticular formation, enable a very rapid turn of the head for the purpose of gaze detection (HASSLER, 1956).

The close connection between the motor function of the eye and head at the reticularis level is also supported by the cervical nystagmus, which is observed in cervical syndrome. For this detailed studies by BARANY exists (1918), FRENZEL (1923), PHILIPSZOON (1962), ANDERSON (1977) and NORRE (1979). The posterior roots C1 to C3 play an important role in the transmission of proprioceptive stimuli from the craniocervical region; they represent the “counterpart” of the accessory nerve (FITZGERALD et al., 1982).

In summary, it can be stated that the formatio reticularis in the coordination of the senses of balance, in the the control of head motor function and in the corresponding modulation of the skeletal muscle tone occupies a central position (DUENSING and SCHÄFER, 1960, NIEUWENHUYS et al., 1980). However, there is no evidence to support the assumption that the lesion responsible for ST is localized in the reticular formation itself localized.

PART II

3.0 The treatment of ST

Causal treatment of ST is not possible because the etiology and pathogenesis is still unclear. This results in the large number of proposed treatment methods including the wide range of proposed surgical procedures. In addition to psychotherapy and behavioral therapy, a variety of drug treatment approaches have emerged, which I will discuss later. First, I would like to discuss the surgical treatment of ST, as the development of the various surgical methods goes back a long way and essentially took place before the introduction of specific forms of drug treatment.

3.1 Surgical treatment

In principle, the surgical procedures known today for the treatment of ST can be divided into 2 groups:

1. denervating interventions on the motor terminal pathway.
2. stereotactic (functional) interventions on the central nervus sytem (CNS).

In chronological order, I would like to start with the purely symptomatic, destructive and denervating surgical methods.

3.1.1 Denervating interventions on the motor terminal pathway

3.1.1.1 Myotomy

As far back as the year 1641 reached the attempts of MINNIUS (cited by STEYERTHAL, 1906) to influence the torticollis-symptoms by severing individual neck muscles. Later myotomies, especially of the SCM muscle, were also reported by MIKULICZ (1895), de QUERVAIN (1896), KOCHER (1912) and MANN (1921). However, this method was soon abandoned as

denervation of the muscles involved in the ST proved to be more effective and longer lasting. However, the transection and resection of individual neck muscles for the treatment of ST was again proposed in a more recent publication from the People's Republic of China (XINKANG, 1981).

3.1.1.2 Extraspinal procedures (neurotomy)

In 1834, BUJALSKI (cited by WYCIS and MOORE, 1954) described the extraspinal transection of the accessory nerve, which is not sufficient as the sole measure.

The transection of the 4 upper cervical nerves was first performed by 1888 by GARDNER. Of 15 such operations 11 healings and 4 improvements are noted (citation: VOELKER in: GARRE, KÜTTNER, LEXER (eds.) Handbuch der praktischen Chirurgie (Handbook Practical surgery) (1930).

In 1891, KEEN first cut, apart from the accessory nerve, only on one side the posterior branches of the three upper cervical nerves. FINNEY and HUGHSON (1925) recommended bilateral extraspinal transection of the posterior branches of the 3 upper cervical nerves together with bilateral transection of the accessory nerve. After initially only slight improvement, they observed a decisive regression of symptoms only 6 months to 3 years after the operation. Similar operations were also performed by COLEMANN (1927) and others.

3.1.1.3.1 Intraspinal cervical rhizotomy

3.1.1.3.1.1 Rhizotomy of cervical posterior roots

The first intradural procedure based on FOERSTER's operation for LITTLE's disease was reported by TAYLOR (1915). To influence the spasticity, he severed the posterior roots of the four upper cervical nerves. The same procedure was later recommended by FRAZIER (1930), who published four cases operated on in this way in which the torticollis symptoms had regressed within a few months. He also referred to FOERSTER

(1918), who recommended dorsal rhizotomy for the treatment of spastic paralysis. In this regard PATTERSON and LITTLE (1943) found that although ST symptoms improved immediately after severing the upper cervical posterior roots alone, recurrence occurred in most cases.

The dorsal rhizotomy is based on the generally accepted view of SHERRINGTON (1907, quoting FRAZIER, 1930) that the muscle tone is controlled by the muscle spindle afferents and the posterior roots.

3.1.1.3.1.2 Rhizotomy of cervical anterior roots

In 1920, FOERSTER first described the intraspinal, intradural transection of the 4 upper anterior and posterior cervical roots for the treatment of ST. In 1924, McKENZIE published a case operated on by CUSHING in which the motor and sensory roots of the 1st, 2nd and 3rd cervical nerves were cut on one side. In addition, the SANR was dissected shortly before its entry into the jugular foramen. The patient recovered well from the procedure and was largely symptom-free 6 weeks after the operation. DANDY (1930) took up FOERSTER'S procedure and modified it: He only cut the 3 upper motor cervical roots on both sides intradurally, as well as both accessory nerves peripherally on the neck. More or less modified this surgical procedure was also used by OLIVECRONA (1931), KAPPIS (1934), TÖNNIS (1934, 1935), WYCIS and MOORE (1954), McKENZIE (1955), ARSENI and SANDOR (1960), SÖRENSEN and HAMBY (1965), KRAYENBÜHL and YASARGIL (1965) and WYCIS and GILDENBERG (1965).

The aim of rhizotomy and neurotomy is the deafferentation of the neck muscles involved in the symptoms (SCHALTENBRAND, 1935). These are exclusively non-reversible, destructive interventions. In addition, these operations were associated with a considerable surgical risk, at least in the early years. As late as 1979, SCOVILLE and BETTIS warned of fatal complications with bilateral infarction of the medulla oblongata caused by injury to small root arteries. They therefore recommended, as did FABINYI and DUTTON (1980) and

COLBASSANI and WOOD (1986), a microsurgical procedure. 11 lethal complications in a total of 277 operations were reported (PUTNAM and GLASER, 1949, SORENSEN and HAMBY, 1965, HAMBY and SCHIFFER, 1969, ARSENI and SANDOR, 1959, ARSENI and MARETSIS, 1971, SCOVILLE and BETTIS, 1979, ADAMS, 1984).

Further disadvantages of the method after unilateral or bilateral cutting of the C4 root are the partial or complete paralysis of the diaphragm and the not insignificant considerable limitation of voluntary head motor skills, combined with instability of the head posture. In many cases patients are dependent on stabilization of the head with a cervical collar after such an operation. In addition, HAMBY and SCHIFFER (1969) observed postoperative swallowing difficulties in 30% of cases, which were mostly of a temporary nature.

3.1.1.3.1.3 Results of cervical rhizotomy

The literature shows that to eliminate the torticollis, bilateral transection of the ventral roots roots C1-C3 and bilateral intracranial transection of the accessory nerve at level C1 is necessary (BRÄUTIGAM, 1954). TASKER (1976), on the other hand, suggested the peripheral transection of the muscle branches of the accessory nerve, since the nerve supply of the SCM and trapezius muscles varied. Therefore, with exclusively intracranial transection of the SANRs a complete denervation of the accessory nerve supplied muscels may not be achieved.

In DT, the results of rhizotomy are poor, so that stereotactic treatment is recommended in these cases (ARSENI and MARETSIS, 1971). In the “pure” ST cases, the compilation of various published series gives the following picture: According to the authors, out of 277 patients operated on authors, 209 (75%) showed a very good to good result. In 57 patients (21%) the symptoms remained unchanged or worsened. 11 patients (4%) died as a result of the procedure (PUTNAM and GLASER, 1949, McKENZIE, 1955, SORENSEN and HAMBY, 1965, HAMBY and SCHIFFER, 1969, WYCIS and GILDENBERG, 1965, ARSENI

and MARETSIS, 1971, SCOVILLE and BETTIS, 1979, KROO et al., 1979, HAYWARD, 1986).

It must be added that after cervical rhizotomies have been performed, in many cases additional extraspinal severing of the accessory nerve and the motor branches of the cervical nerves were necessary to achieve a satisfactory result.

It is also noticeable that the results of the series published since 1979 were significantly worse. For example, SCOVILLE and BETTIS (1979) reported an improvement of symptoms in only 62% of their 23 cases, KROO et al. (1979) in 66% of their 12 cases. HAYWARD (1976) reported only 6 (40%) improvements in his 15 cases.

SCOVILLE and BETTIS (1979) therefore recommended the following surgical strategy: In the case of purely horizontal ST, only the accessory nerve in the neck should be severed initially. If the symptoms do not improve, a bilateral vertebral angiogram should be performed and only if this shows normal vascular conditions should the decision be made to perform a rhizotomy. To ensure adequate postoperative support of the head, the patient is fitted with a neck collar. In 1976, TASKER wrote: "At the present time, I consider that denervating surgery is be the best we have to offer; the palliative procedure leads in most patients patients leads to a longer-lasting improvement in symptoms with low surgical risk".

3.1.1.4.1 Selective peripheral denervation

As late as 1976, BERTRAND recommended stereotactic interventions for the treatment of ST (improvement in 70% of cases), as the greatest disadvantage of cervical rhizotomy was the resulting instability of the head balance.

Just 2 years later, BERTRAND et al. (1978) reported the first positive experiences with a combined stereotactic and peripheral surgical procedure for the treatment of ST. Now they were based on the observation that after uni- or

bilateral thalamotomy of a mixed patients with ST and DT, satisfactory results were achieved in only about 50% of cases.

After bilateral thalamotomy, they saw only 4 patients with 2 good results, in 9 patients with unilateral thalamotomy only 5 satisfactory results. For this reason, they also performed a modified form of peripheral denervation of the posterior branches from C1 to C4, as described by KEEN (1891), in 5 patients who still had considerable residual symptoms. This ultimately resulted in a good regression of the torticollis symptoms.

A few years later, BERTRAND et al. (1981, 1982) completely changed their surgical approach to ST, as they after local anesthetic blockage of nerves supplying the cervical muscle during the EMG examination, they observed a clear to complete improvement of the torticollis symptoms. They now assumed to make a distinction between DT and ST and distanced themselves from the stereotactic treatment of "pure" ST, which today is the most frequently performed surgical method worldwide. BERTRAND et al. emphasized that stereotactic surgery should only be reserved for the more severe forms of torsion dystonia. They proposed a new surgical method for the treatment of ST: The "selective peripheral denervation".

The most important prerequisite for the successful of the procedure is the careful electromyographic analysis of the individual neck muscles involved in the movement disorder. The authors emphasized that sometimes unsuspected muscles, especially the agonists of the contralateral side may be involved.

3.1.1.4.2 Description of the method

The neck muscles most strongly involved in ST symptoms are are blocked with lidocaine (1%) in order to identify the less active muscles. The patient is also given an impression of the effect of permanent denervation. The final surgical denervation is performed under light anesthesia without

muscle relaxants to allow intraoperative stimulation of the motor nerve branches. The posterior branches from C1 to C6, and sometimes also C7, are exposed one after the other on both sides via a lateral approach. With the help of stimulation, the posterior ramus C1 is sought out microsurgically, followed centrally and severed at the level of the posterior arch of the atlas. The posterior branches of C2, C3 and C4 are also sought out and severed. In contrast, the posterior branches of C5, C6 and occasionally C7 are only resected on one side, depending on the EMG findings. Finally, the branches which supply the SCM muscle are cut, while the branch leading to the trapezius muscle is spared. Finally, the complete denervation of the individual muscles is checked by high-voltage stimulation.

3.1.1.4.3 Results of selective peripheral denervation

In the meantime, BERTRAND and MOLINA-NEGRO (1987) have published a study on 111 patients who underwent surgery. 97 patients (87%) were satisfied with the results of the procedure, which was combined with intensive postoperative physiotherapy. In 12 cases, only a slight improvement was seen, while in 3 patients with rotatory torticollis the symptoms remained unaffected. It is determined that despite extensive denervations a good restoration of normal head mobility could be achieved. No complications were observed. The only consequences of the surgical procedure were atrophy of the denervated muscles and a sensory disturbance in the region of the occipital nerves.

3.1.1.4.4 Summary

BERTRAND(1987) lists the following advantages over rhizotomy:

1. Only muscles exclusively involved in the torticollis symptoms are denervated, while the anterior long neck muscle groups remain intact, so that the rehabilitation of the patients is much easier and more complete than after a rhizotomy.

2. Laminectomy is not necessary, so that the stability of the cervical spine is not impaired.

3. In contrast to cervical rhizotomy, denervation can be performed unilaterally down to C7 as the phrenic nerve is spared.

4. The procedure can be individually tailored to the needs of the individual patient.

With regard to point 2, however, it should be noted that after a bilateral denervation of the neck muscles and the SCM muscle, however, late effects in the form of a "Swan neck deformity" must be expected.

3.1.2 Stereotactic ("functional") interventions on the CNS

3.1.2.1 Development of the method

Following fundamental animal experiments (SPIEGEL, 1927, HESS, 1941) stereotactic interventions in humans were first performed in 1947 by by SPIEGEL et al..Due to the importance of the basal ganglia for the modulation and maintenance of skeletal muscle tone, as researched in particular by SPIEGEL, HESS and DENNY-BROWN (1962), this procedure initially was used only in humans to influence athetotic or choreatic movement disorders (MUNDINGER and RIECHERT, 1961, MUNDINGER, 1965, 1969, MUNDINGER et al., 1970, HASSLER and RIECHERT, 1954, MOLINA-NEGRO, 1979, ANDREW, 1981, SCHMIDT and POTTHOFF,1981).

On the basis of the stem-ganglion-theory postulated for the ST and based on the animal experimental experience of HESS (1941) (see p. 30), a patient with a "pure" ST was stereotactically treated for the first time in 1956 by HASSLER and DIECKMANN (1970) in collaboration with MUNDINGER and RIECHERT (1961). A target point was chosen in the medial parts of the oral ventral nuclei of the thalamus. After a relatively good initial result, however, it was necessary to cut additionally the accessory nerve for the hypertrophied

SCM muscle. Even with the following operations, a satisfactory result could not be achieved in this way.

Later, HASSLER and DIECKMANN (1968, 1970) modified their operational procedure: Since 1964, every torticollis patient was initially analysed regarding his misalignment of the head. In patients with purely horizontal symptoms, the subthalamotomy according to MUNDINGER (1965) with interruption of pallido-thalamic fibers, also called kampotomy, was performed. In patients with rotatory torticollis, on the other hand coagulations were performed in the inner section of the oral ventral ventral nuclei of the thalamus. However, it turned out that the rotatory ST was far more therapeutic influenceability lagged far behind the horizontal ST. The most common form of ST, the combined horizontal-rotatory torticollis, was easier to influence, but not as well as the purely horizontal ST.

Further target pots and modifications subsequently were specified for example the pallidotomy of the inner pallidum limb according to TAILERACH et al. (1950, 1954), the thalamotomy according to HASSLER (1956), bilateral coagulations in the nucleus interstitialis Cajal of the mesencephalon and in the fasciculus medialis longitudinalis according to SANO et al.(1970) or combined lesions, with thalamotomy and pallidotomy (LAITINEN, 1963, KRAYENBÜHL and YASARGIL, 1965, LAITINEN and JOHANSSON, 1966, HASSLER and DIECKMANN, 1970, ARSENI and MARETSIS, 1971, VASILESCU and DIECKMANN, 1975, LÜCKING and STRUPPLER, 1977, AMANO et al., 1979, van ESSEN et al., 1980, LOZANO-SAAVEDRA, 1981, HEIKKINEN, 1986) (see Fig. 4).

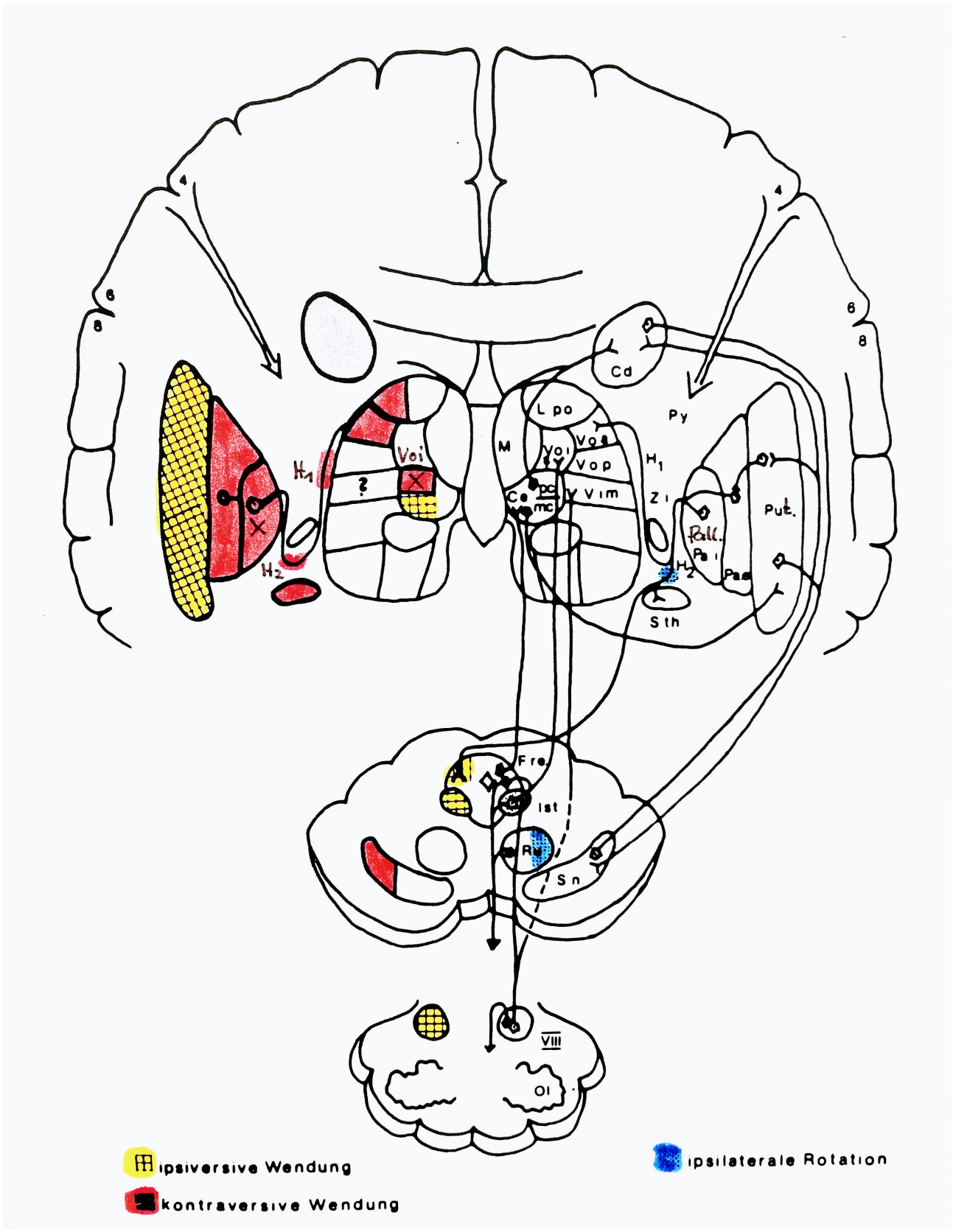


Fig. 4: Target points of stereotactic operations (from THÜMLER: Torticollis spasmodicus (1983)).

3.1.2.2 Results of stereotactic treatment of ST

MUNDINGER et al. (1972) reported satisfactory long-term results in 60% of 52 patients, COLBASSANI and WOOD (1986) in 56% of cases.

In 1976, COOPER published the results of 160 stereotactically operated ST patients. He also achieved satisfactory results in 60% of cases. However, like WYCIS (1970) and ANDREW (1981), he emphasized that he did not see a sufficient effect after unilateral elimination, bilateral lesions of the ventrolateral nucleus in the thalamus were necessary to achieve good results. However, this resulted in a higher surgical risk. He therefore came to the conclusion that a procedure with only a 60% chance of success should not be recommended.

GOLDHAHN and GOLDHAHN (1977) saw in 24 stereotactically operated ST patients 14 times (58%) a satisfactory condition. MOLINA-NEGRO (1979) reported good results in 65% of 54 torticollis cases. VAN ESSEN et al. (1980) found all of their 17 thalamotomized torticollis patients improved.

HERNESNIEMI and LAITINEN (1977) presented a long-term study of 80 ST cases, of which 70 patients had been operated on in various ways. With regard to the success rate for stereotactic surgery, they came to disappointing results. Out of 17 patients, only one showed a good result, only 5 (30%) patients were able to work. Good results in 47% of cases were reported in 1981 by STEJSKAL et al. (1981).

OJEMANN and WARD (1973) recommended, like COLBASSANI and WOOD (1986), the stereotactic approach only for those cases in which the torticollis is a partial symptom of a torticollis dystonia. In contrast to this, GOLDHAHN and GOLDHAHN (1977), like HASSLER and DIECKMANN (1970), found in undifferentiated patient population that the horizontal torticollis was best to influence.

SCHMIDT and POTTHOFF (1981) made the following observation:

"ST, which also belongs to the extrapyramidal movement disorders is far more difficult to influence than all other movement disorders. Whereas in the case of purely horizontal ST, good or satisfactory results can be expected in 50-70% of cases, the success rate for ante- and retrocollis, but especially in mixed forms of the disease decreases significantly".

MÜLLER (1983) wrote: "When looking at the published and own surgical results, there is a surprising discrepancy between torsion dystonia and ST. The long-term results for torsion dystonia with good effect in 50%, moderate in 29%, unchanged in 13% and worsening in 8% are fully confirmed by our own patient collective. With ST, on the other hand, the results vary greatly. On average, good to satisfactory results are reported for two thirds of patients. Other authors, however, have achieved good results in only 50% of cases. Moreover, these results were often only achieved by additional peripheral interventions".

3.1.2.3 Summary

Summarizing the treatment results achieved to date with stereotactic surgery, it can be seen that after initially quite optimistic reports, a sobering up has occurred. Despite 30 years of experience in the stereotactic treatment of ST, experimenting with the most diverse target points and despite often bilateral interventions, the treatment results have not improved at all. On the contrary, many authors who previously advocated stereotactic interventions for ST now have turned to other surgical procedures: GILDENBERG (1980) now favors spinal cord stimulation for the treatment of ST. BERTRAND (1981) developed the peripheral selective denervation and DIECKMANN (1981), as well as MUNDINGER (1977) experimented with deep brain stimulation. Today, DIECMANN (1985), like NITTNER (1986), recommends epidural spinal cord stimulation and peripheral selective denervation.

Stereotactic brain surgery should therefore only be used for central motor movement disorders, including torticollis

dystonicus, whereas ST should be treated with peripherally applied surgical methods (KRAYENBÜHL and YASARGIL, 1965, ARSENI and MARETSIS, 1971, OJEMANN and WARD, 1973, COLBASSANI and WOOD, 1986).

3.1.3 Deep brain stimulation (thalamic - subthalamic)

In 1977, MUNDINGER, who had extensive experience in the stereotactic treatment of ST patients, introduced a new stereotactic, functional treatment method of ST with deep brain stimulators. He reported on 7 patients, in whom permanent implantation of a brain stimulation system into the thalamic nuclei or the subthalamic zona incerta involving the H1 and H2 bundles an improvement was achieved. A stimulation lasting 30 to 40 minutes, which was determined and carried out by the patients themselves, resulted in control over head position and mobility for a period of up to 7 hours with elimination of the symptoms of ST. MUNDINGER justified the new method because stereotactic operations, sometimes in combination with peripheral interventions, have led to the best results to date in around 60% of patients, although some patients experience recurrences after a more or less long pain-free interval and there is an increasing complication rate, especially after bilateral interventions.

Long-term studies on the results with this procedure are not yet available. The literature contains hardly any other reports or experience with deep brain stimulation. Only ANDY (1983) reported good results with thalamic stimulation for the treatment of movement disorders. Out of 8 patients only one suffered from DT, another from "torticollis".

3.1.4 Cervical dorsal spinal-cord stimulation (DCS)

GILDENBERG (1977, 1979) presented a further, non-destructive, less invasive method for the treatment of ST: The high DCS.

3.1.4.1 Description of the method

At the level of the 5th or 6th cervical vertebral spinous

process, in local anesthesia, a hollow needle is inserted diagonally upwards into the into the epidural space. An electrode is inserted through the hollow needle and, under image converter control cranially to the level of the lower edge of the posterior arch of the atlas and placed there. The indifferent electrode is placed on the skin of the neck. With a small stimulation device, the high cervical medulla now can be stimulated. Frequencies between 1100 and 1400 Hz are preferred.

The method introduced by GILDENBERG (1977) is based on the idea that stimulation generates additional afferent stimuli in the neck region, which by influencing the tonus-regulating systems lead to an altered motor response. It is assumed that high-frequency stimuli block or depolarize the proprioreceptive fibres that are involved which play a role in the regulation of tonic neck reflexes. Later, GILDENBERG (1979) first introduced the transcutaneous cervical (high) DCS in order to draw attention to the possible effect of an implanted stimulation system. With 23 patients tested in this way GILDENBERG decided to implant the system in 8 cases.

3.1.4.2 Results of the cervical DCS

In the first 6 ST patients treated by GILDENBERG, one very good and three good results were achieved. Another patient with a torsion dystonia did not experience any improvement by this method.

In 1985, WALTZ et al. and DIECKMANN and VERAS published their experiences with DCS. WALTZ et al. (1985) reported on their observations of 63 patients with permanently implanted systems. In 23 patients (36%) they found a very good improvement of the symptoms, in 20 patients (32%) patients (32%) showed a slight improvement. DIECKMANN (1985) found a clear improvement in torticollis symptoms in half of his 18 patients. In 28% of the cases he still speaks of a satisfactory result. If both patient series with a total of 81 patients are combined, the result is satisfactory in 68% of cases. We ourselves have used this method on one patient

without success.

REYNOLDS and SHETTER (1983) reported an unusual complication. After implantation of a stimulation system, a spastic tetraparesis occurred. A scar had formed on the epidural electrode, which led to compression of the cervical cord.

The few publications to date on this treatment technique do not yet allow a conclusive assessment of this non-destructive and only slightly invasive method.

3.1.5 Leukotomy - cortectomy - chordotomy

For the sake of completeness, at the end of the discussion of surgical ST treatment methods, some open cerebral interventions rarely used in the early 1940s should be mentioned. SCHÜRMANN (1953): "In a series of cortectomies, KLEMME also reported an improvement of symptoms in torticollis, but DAVID and coworkers saw in one of their cases, where a torticollis spasticus was added to the torsion spasm of the arm, that the last symptom disappeared after removal of areas 4 and 6., but returned after only 4 1/2 months. MEYER experienced a complete failure with subcortical cutting of the pallidofugal fibers in a case of torticollis. Likewise unconvincing and unsuccessful were attempts to treat spasmodic torticollis with the prefrontal lobotomy by WOHLFAHRT and FORSOCK, GIRARD, BORDET and DEVOC." In the same paper SCHÜRMANN (1953) also reported on his own attempts to influence the ST in three cases by means of a cervical "extrapyramidotomy", which, however, failed completely.

3.2 Conservative treatment of ST

Due to the relatively uncertain prospects of success and the destructive nature of surgical treatment procedures, conservative treatment of ST should be the first choice of possible treatment methods. In most patients, especially if the Symptoms are only mild, conservative treatment measures can alleviate the symptoms. This applies in particular to the special torticollis gymnastics, exercise baths, but also

other physical therapeutic measures such as massages and heat applications. With the help of physiotherapy exercises in particular, the patient learns to train the antagonistic muscles, to bring the head to a central position and hold it.

3.2.1 Non-invasive treatment methods

3.2.1.1 Physiotherapy

Non-invasive treatment methods for ST include general physiotherapy, special torticollis-gymnastics, exercise baths and swimming, local heat applications, massages and, to a certain extent, chiropractic measures. In addition to general physiotherapy, in our experience the specific symptoms of torticollis play an important role in the overall treatment concept, especially after previous operations. The treatment techniques according to BRUNKOW and VOIJTA should be mentioned here (DIECKMANN, 1977, HADANK, 1981). In a study we carried out in 1983 on the effectiveness of non-invasive treatment methods, it was found that special torticollis gymnastics produced the best treatment results. Of 90 ST patients surveyed, 58% reported an improvement in symptoms and relief of discomfort. Deteriorations were not described under torticollis gymnastics (HAGENAH 1983).

Exercise baths and swimming were perceived by 54% of patients as pleasant and pain-relieving. Here, however worsening of the symptoms were reported in 8% of patients. In 3rd place in terms of the effectiveness of conservative treatment methods is the general physiotherapy with improvements in 47% of cases. In further descending order are local heat applications with 33%, massages with 30%, local injections into the neck muscles with 21%, chiropractic measures with 10% and acupuncture with with 9% improvement. Cervical spine stretching led to alleviation of symptoms in 11% of cases, but also in 26% of cases led to worsening. With the highest rate of worsening of all conservative treatment measures, cervical spine stretching should be rejected as a treatment method for ST (HAGENAH et al. 1983, HAGENAH, 1983).

3.2.1.2 Sensory biofeedback therapy

Biofeedback therapy is a conservative treatment method that aims to teach the patient controlled activation and relaxation of pathologically innervated muscles. The individual methods used for feedback therapy vary. Biofeedback is used when inner-organ processes that are normally hardly accessible to self-observation are made perceptible through visual or acoustic feedback. The aim of the feedback process is to bring the physiological reaction under voluntary control. Experiments have shown that the innervation rate of individual motor units in the skeletal musculature can be fed back and changed arbitrarily (DAHME, 1980).

At the beginning of the 1970s, this discovery led to a "biofeedback boom", especially in the USA, which is comparable to the acupuncture wave. The aim of feedback therapy in ST is to enable patients to voluntarily control and direct the pathologically innervated muscles. Patients receive a visual or acoustic signal that gives them feedback on the current activity in the affected muscles. The muscle activity is recorded by EMG (CLEELAND, 1973, BRUDNY et al., 1974, ROXANAS et al., 1978, KEEFE and SURWID, 1978, GERBER et al., 1983, LEPLOW et al., 1983).

BRUDNY et al. (1974) found that feedback therapy still had a favorable influence on ST in 45% of patients after a 4-year follow-up. In our own patient population, we saw a temporary improvement in 3 out of 6 patients treated. The feedback therapy appears to have a positive influence on ST symptoms in some patients, at least temporarily (LEPLOW et al., 1983).

3.2.2 Drug treatment of ST

Apart from a few publications that report on the drug treatment of ST with substances whose mechanism of action is not known, such as treatment with lithium carbonate (FOERSTER and REGLI, 1977, SACHDEV and BRODSKY, 1979), the drugs used can be categorized according to six principles of action:

1. centrally acting muscle relaxants,

2. tranquilizers,

3. neuroleptics,

4. anticonvulsants,

5. anticholinergics and

6. dopaminergics

The literature also contains reports on the treatment of ST with drug combinations and rarely used pharmacological substances. In particular mention should be made intramuscular injections of

a) local anesthetics and

b) botulinum toxin.

3.2.2.1 Muscle relaxants

Treatment with centrally acting muscle relaxants alone (Sirdalud (R) (tizanidine), Musaril (R) (tetrazepam), Muskel Trancopal (R) (chlormezanone)) has been shown to be largely ineffective in ST (HAGENAH, 1983, ten HOUTEN, 1984).

3.2.2.2 Tranquilizers, antidepressants

Treatment with tranquilizers (Valium (R) (diazepam), Tranxilium (R) (dipotassium clorazepate), Tavor (R) (lorazepam), Lexotanil (R) (bromazepam), L-tryptophan, etc.) give good results in a favorable picture for diazepam in particular (LAL et al. 1979, PETELIN et al. 1980, LAL, 1981, DISERTORI et al., 1982, FRANCIS, 1983). In this group we saw a positive effect in 46% of the patients, deterioration in 15% of the patients (HAGENAH, 1983, ten HOUTEN, 1984).

3.2.2.3 Neuroleptics

The group of neuroleptics includes Dartal (R)(thiopropazate), Haldol (R) (haloperidol), Tiapridex (R)(benzamide derivative) and Decentan (R) (perphenacin). Of these drugs, Dartal (R) showed the best effect with 36% improvement and 7% worsening. 14% of patients reported side effects such as tiredness and fatigue, but these were usually improved by the additional administration of anticholinergics (HAGENAH, 1983).

Haldol (R) is frequently prescribed, especially in combination with L-dopa preparations (SHAW, et al. 1972, DOMZAL, 1978, LAL, 1979, LANG et al, 1983, GILBERT, 1972, BIGWOOD, 1972, COUCH, 1976, HAGENAH, 1983). About one third of the patients patients reported improvements.

Tiapridex (R) and Decentan (R) proved to be effective according to our experience, which is in line with the literature, proved to be largely ineffective (DIVISIA and GIRARD-MADOUX, 1978).

3.2.2.4 Anticonvulsants

Treatment with anticonvulsants, such as phenytoin, carbamazepine and clonazepam, has also been tried at high doses without success (BERTRAND, 1978).

3.2.2.5 Anticholinergics

Anticholinergics are another group of preparations that with a target on the neurotransmitter system of the basal ganglia are used. In smaller patient cohorts, positive effects on the symptoms of torticollis have been described (FOLTZ et al., 1959, DOMZAL, 1978, LAL, 1979, SCHEROKMAN et al., 1986). LANG et al. (1983) contradicted these reports, they could not have a stronger effect with the classical anticholinergic atropine than with a placebo.

In the patient population we studied, Akineton (R) (biperiden), with a similarly favorable effect as with Dartal

(R), but with somewhat more frequent side effects such as dry mouth. Artane (R) (trihexyphenidyl), Tremarit (R) (metixen HCl) and Sormodren (R) (bornaprin HCl) proved to be less effective (HAGENAH, 1983).

3.2.2.6 Dopaminergics

Most publications on the drug therapy of ST are found for the treatment analogous to Parkinson's syndrome with dopaminergics (BIGWOOD, 1972, SCOVILLE and BETTIS, 1979, WEST, 1977, GILBERT, 1971, 1972, HAGENAH, 1983, HIRSCHMANN and MAYER, 1964, COLBASSANI and WOOD, 1986, ANSARI et al., 1972, TOLOSA, 1978, DOMZAL, 1978, LAL et al., 1979, PETELIN et al., 1980, OLANOW, 1981, LEENDERS et al., 1985, LANG et al., 1983).

Apart from GILBERT (1971), the ineffectiveness of this treatment was unanimously was noted by all other authors. Where positive results were seen, possibly the combination with Haldol (R) had the the main part of the therapeutic success. In our own patient population we did not see any positive effect with Madopar (R) (L-dopa) or PK Merz (R) (amantadine sulphate), but rather a deterioration. In summary, therapy with these preparations does not appear to make sense.

3.2.2.7 Lithium carbonate

There have been isolated reports on the treatment of ST with lithium carbonate (COUPER-SMARTT, 1973, FOERSTER and REGLI, 1977, SACHDEV and BRODSKY, 1979, LIPPMANN and KAREUS, 1983). The mechanism of this treatment is not known, with a total of 4 published cases no assessment can be made. There is no personal experience with lithium therapy.

3.2.2.8 Rarely used medications

Some authors reported on the treatment of ST with the dopamine agonists and prolactin inhibitors Pravidel (R) (bromocriptine) (JUNTUNEN, 1979) and Dopergin (R) (lisuride)

(BASSI et al., 1982), the Roborans L-glutamine (KOREIN et al., 1981), and with nitric oxide, for which GILLMAN and SANDYK (1985) observed a favorable effect on ST symptoms. Nitric oxide in analgesic concentrations is said to have an influence on the endogenous opioid system and to stimulate it.

For these drugs too, due to the small number of published cases, an assessment of the treatment results is not possible.

3.2.2.9 Drug combinations

Combinations of medications are often used in therapy, most frequently, as already mentioned, dopaminergic substances together with haldoperidol or diazepam.

3.2.2.10 Intramuscular injection of local anesthetics

SCHALTENBRAND (1935) for the treatment of ST proposed extensive, systematic injections of novocaine into the entire cervical musculature. The aim of the treatment was to achieve prolonged neural atrophy and deafferentation of the neck muscles. He practically cured 4 patients with this method. We were also able to achieve a temporary interruption of the ST symptoms in some cases with unilateral, local injections of bupivacaine (R) in the vicinity of the upper cervical joints, resulting in a certain deafferentation of C1 and C2.

3.2.2.11 Intramuscular injection of botulinum toxin

Recently, studies have been published that report good results with the local application of botulinum toxin. The working group around TSUI (1985, 1986) injected doses of botulinum toxin intramuscularly into the the neck muscles involved in torticollis symptoms, resulting in paralysis of the corresponding muscles. In 19 patients treated in this way, 18 improvements were observed. The positive effect of the venom has already been described for the treatment of hemifacial spasm and blepharospasm. Here, further development

will show whether the efficiency of this method will be confirmed.

3.2.3 Summary of conservative treatment methods

Conservative treatment of ST, also in conjunction with with surgical treatment, is an essential part of the overall treatment concept for this disease. In many cases it leads to a reasonably satisfactory treatment outcome for both patient and doctor. This is particularly true for the majority of mild ST cases. Among of all possible treatment methods, surgical therapy should be seen as the "ultima ratio". It should be only be considered if, in the case of longstanding, severe and painful torticollis symptoms, all conservative treatment measures have not led to any improvement and the suffering has reached an unbearable level.

Any therapy should start with physiotherapy and swimming. If this does not bring about sufficient improvement, the next measure should be additional bio-feedback training should also be carried out. Only if the ST symptoms cannot be influenced with these measures medical treatment should be attempted. Attention must be paid to possible side effects of the medication. In our experience, therapy with Dartal (R), possibly in combination with the membrane-stabilizing drug Xylotocan (R), appears to be the most suitable. Addiction problems did not occur (HAGENAH, 1983).

Prolonged treatment with Valium (R) or Musaril (R) is not recommended because of the risk of addiction. It should only be used for a limited period of time for painful, cramp-like muscle tension in the shoulder and neck area. The special ST gymnastics according to VOIJTA/HADANK should be continued alongside drug treatment without fail.

Treatment with substances that work on the neurotransmitter system of the basal ganglia and the corresponding extrapyramidal pathways did not have the desired effect. This fact can be interpreted as a further indication that a primary involvement of the basal ganglia in the pathogenesis

of ST is probably not present (HAGENAH et al. 1983).

3.3 Critical evaluation of all ST treatment methods

In a critical assessment of all the treatment methods mentioned so far, the following points should be emphasized:

1. physical therapy has a favorable effect on the symptoms of ST. In addition to surgical treatment, physiotherapy and swimming are the most important measures in the treatment of ST.

2. Drug treatment of ST only leads to improvement in about one third of patients. It is also associated with side effects, some of which are significant.

3. All conservative treatment options should be exhausted before considering surgical treatment.

4. while stereotactic interventions for extrapyramidal extrapyramidal movement disorders lead to relatively good results, the results in the treatment of ST can be described as poor. If a destructive intervention is accepted, improvement can only be expected in about 40 to 60% of cases. The possible side effects, primarily hemiparesis and speech disorders, are severe, irreversible and occur in up to 50% of cases.

5. The results of rhizotomy, which is a peripheral but destructive procedure, are better in ST compared to torsion dystonia. However paralysis of large parts of the neck musculature must be accepted. The voluntary mobility of the head is considerably restricted. The patient must learn to balance the head on the cervical spine. In addition to the risk of serious surgical complications, the secondary consequential damage with degenerative changes to the cervical spine must also be taken into account with this method.

6. peripheral selective denervation appears to be a promising procedure for the treatment of ST, but it is

also one of the destructive interventions. In special cases, this operation can be of benefit.

It must be emphasized once again that all methods available to date for the treatment of ST, whether conservative or surgical, are not causally effective and are therefore ultimately unsatisfactory. It often takes many attempts with the individual patient to find the most favorable treatment method. In our experience with over 300 ambulant Torticollis patients, a satisfactory treatment result can be achieved by conservative treatment in 85% of cases, but in the remaining severe ST cases, where conservative therapy fails, surgical measures must be considered.

PART III

4.0 The "neurogenic ST"

In 1979, after a critical review of our treatment results for ST, we realized that the stereotactic interventions had led to very poor results. Only 5 of 13 patients could be improved, 8 patients remained unaffected in their symptoms. The rate of side effects (hemiparesis and speech disorders) was 54%. In view of this situation, we could no longer support the basal ganglia hypothesis and the therapy of ST derived from it.

As a result, it became necessary for us to look for new ways in the therapy of ST, especially in the surgical treatment, and to make further efforts to research the etiology and pathogenesis of ST, in order to offer severely afflicted ST patients in particular a more favorable perspective for the prognosis of their disease. Stereotactic surgery was reserved only for cases with DT, while a new surgical approach for ST had to be found, as cervical rhizotomy was not a satisfactory alternative.

4.1 Theoretical principles

4.1.1 The importance of vascular contacts at the entry and exit zones of the cranial nerves

JANNETTA (1977, 1980) on the basis of previous observations by DANDY (1938) and GARDNER (1962), pointed out the importance of neurovascular contacts at the root entry and and exit zones of various cranial nerves, including the facial nerve for the genesis of hemifacial spasm (HFS).

Already 1962, GARDNER and SAVA developed a new hypothesis for the pathogenesis of HFS, taking into account earlier experimental work by UEXKÜLL (1894), GRANIT et al. (1944), FRANKENHAEUSER and NYSTRÖM (1954) and MARRAZZI and LORENTE (1944) on interactions between neighboring myelin-coated

fibers in damaged nerves (ephapse = artificial synapse).

GARDNER (1962) suspected a connection between HFS and aberrant vascular loops in the cerebellopontine angle (CPA) with contact to the facial nerve directly at its exit from the pons. GARDNER wrote: "As a result of my observations, it must be assumed that HFS is an expression of a reversible pathophysiological condition caused by mild, prolonged compression of the facial nerve in the CPA. The HFS can result from a transaxonal "short circuit" on the basis of pressure atrophy of insulating myelin sheaths. The involuntary contractions of the mimic musculature occurring in HFS could be seen as an interaction between the efferent nerve fibers. HFS is a self-sustaining form of synkinesis, with the character of an echoing short circuit of communicating efferent and afferent axons within the afferent axons within the facial nerve root".

JANNETTA (1976, 1980) not only was able to substantiate this hypothesis on the basis of a large, successfully operated patient collective but was also able to substantiate the same pathogenetic basis also for trigeminal neuralgia (TN), glossopharyngeal neuralgia and vagus neuralgia. Today's worldwide experience with microvascular decompression for the treatment of cranial nerve dysfunction syndromes (CDS) has led to a complete rethink for these clinical pictures. HFS and TN, also their etiology and pathogenesis for a long time were still controversial and, until a few years ago, centrally localized lesions also were assumed.

4.1.2 Focal demyelination of cranial nerve roots

Experimentally, BURCHIEL (1980, 1981) was able to demonstrate abnormal impulse generation in focally demyelinated trigeminal nerve roots in monkeys. The transition of the trigeminal root from the brain part to the peripheral nerve was identified as a typical lesion site. This is the border between the central oligodendrocyte segment and the beginning of the SCHWANN-sheaths surrounding the axons in the area of the nerve root exit zone. The cranial nerves are particularly

vulnerable at this junction. Obviously, with the appropriate disposition, pulsating vascular contact is sufficient to cause focal demyelination of the nerve and thus produce an ephapse.

Clinical experience shows that the removal of the vascular nerve contacts without any destructive procedure is sufficient to heal the HFS or TN (JANNETTA, 1980, FRECKMANN et al., 1981, WINKLER et al., 1986).

4.1.3 Working hypothesis of "neurogenic" ST

An analogous approach to microvascular decompression in HFS to the aetiology, pathogenesis and therapy of ST in the literature I could not found. In FOERSTER (1929), however, the following note can be read: "I have found a pronounced torticollis in an aneurysm of the vertebral artery, which exerts a direct stimulus on the intradural section of the accessory nerve and to the first cervical root. More frequently abnormally increased activity of the cervical rotators due to pathological stimulation of the afferent pathways or the sensory receptors of the corresponding muscles themselves, but also other substrates of the neck area. In all these cases, torticollis is caused by reflexes, e.g. irritative lesions of sensitive cervical nerves."

Encouraged by our own experience with microvascular decompression for the treatment of CDS, especially HFS, we asked ourselves whether a peripheral triggering of the disease could possibly also underlie ST, since a certain similarity between HFS and ST cannot be denied. In 1979, we therefore began to look for indications of a peripheral cause of ST, localized outside the CNS. EMG-examinations initially seemed to be a suitable method for this.

4.2 EMG-examinations in torticollis

Like most of the other investigators, our neurophysiologists focused their attention on the EMG of torticollis patients until 1979 on the differentiation between torticollis

possibly caused by psychogenic or central motor factors. In particular, tremor groups and discharge bursts of agonists and antagonists were searched for and, if necessary, analyzed. The rhythmicity and frequency of any tremor, the constancy of the tremor groups and the alternating or synergistic action of agonists and antagonists were used as criteria for the choice of the stereotactic target point (Table 1.) (HAGENAH et al., 1980). It was striking that in pure ST only agonists are involved in the symptoms, whereas in DT antagonistic muscle groups are also involved in most cases (HERZ and HOEFER, 1945, PODIVINSKY, 1968, FASSHAUER, 1980, LÜCKING,1980, BERTRAND,1987).

Table 1: Tremor signs in EMG in torticollis patients (HAGENAH et al., 1981)

n=61	POSITIVE	NEGATIVE	QUESTIONABLE
	29	26	6

Since the beginning of 1980, we have modified the electromyographic technique in torticollis patients in order to also assess potentials of individual motor units. EMG analysis, which is usually used for peripheral neurogenic lesions, however, is complicated in torticollis by the fact that the rhythmic or burst-like discharges hardly allow an assessment of individual potentials and a lateral comparison often is not possible due to the mostly unilateral muscle hypertrophy (HAGENAH et al., 1980, 1981, 1983).

4.2.1 Method

The EMG recording of the individual neck muscles was performed with coaxial needle electrodes. The potentials were displayed optically and acoustically using Medelec-electromyographs. Via integrated thermal recorders the potentials were recorded "online" or as "playback" on coregistering cassette devices for later analysis. Predominantly it was derived from the sternocleidomastoid and

trapezius muscles, less regularly from the splenius capitis muscle and from the short head and neck muscles.

The aim was to visualize and measure individual potentials sufficiently often. During the evaluation it had to be taken into account that some of the derived muscles were clearly hypertrophied. Because of the involuntary partly tonic, partly clonic innervations, it was necessary to adjust to each patient individually. The initially occurring affective increase of the involuntary muscle contractions had to be overcome. It was advantageous if the examiner tried to control the innervation density himself with tactile stimuli.

In this way, in addition to many inconspicuous potentials, pathologically altered motor units could be derived and recorded for a sufficiently long time. The assessment of these potentials was based on the criteria of potential size and width, as well as discharge frequency. With simultaneous registration of a maximum of 4 different potentials in 100 msec. damage was assumed if one of the potentials, often with often over 5 mV, appeared 2-3 times on the cathode ray oscillograph which corresponds to a discharge frequency of 20-40/sec. In addition to the generally known criteria, the discharge frequency must be taken into account because between minimal innervation and involuntary innervations in ST patients there is hardly any possibility of adjustment.

4.2.2 Results

In very time-consuming examinations, we were able to find in two thirds of the torticollis patients examined, reproducibly altered individual potentials that can be regarded as neurogenically damaged. The extent of this damage was, however, not comparable to that of a common peripheral nerve lesion, such as a root compression syndrome. While in peripheral nerve lesions, impulse series of relatively constant duration and frequency can be derived, in the ST were found "impulse series" with inconstant pulse density, irregular duration and uneven frequency. These signs were often found in clinically normal or even hypertrophied

muscles. A distinction as to whether this "neurogenic damage" was of a primary or secondary in nature, i.e. only after the torticollis symptoms were present was not possible. Since such findings in patients with DT were not found, we came to the conclusion that these findings could be an expression of an accessory nerve irritation (Tab. 2.) (HAGENAH et al., 1980, 1981, 1983).

Table 2: Neurogenic lesions according to EMG findings (HAGENAH et al., 1981)

n=32	FRESHER	OLDER	OLD
	11	23	22

4.3 Anatomical studies

Further evidence for a peripheral triggering of the ST we expected from an anatomical study of the topographical conditions in the area of the craniocervical junction (KOSAK et al., 1981, HAGENAH et al., 1983).

4.3.1 Material and method

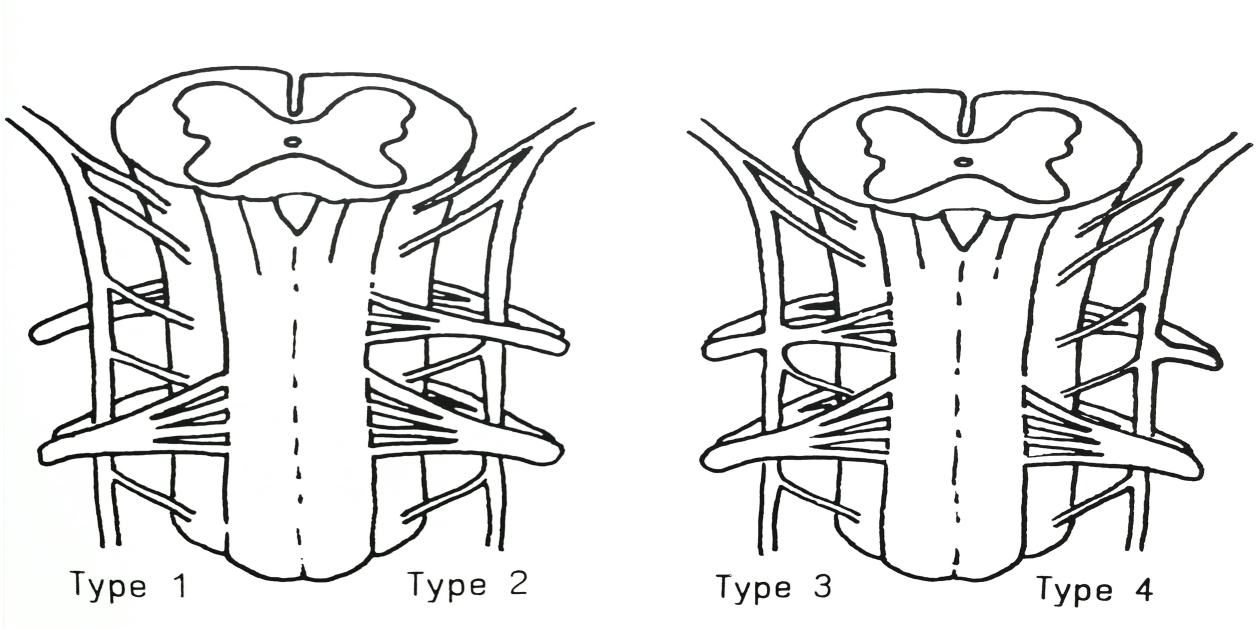
The craniocervical region was dissected from 50 cadavers from the general dissection material and the SANR from its origins to its emergence from the posterior skull base on both sides. After macrophotographic documentation, under loupe magnification, 100 SANRs and their topography were examined. Attention was also paid to variations of the fila radicularia dorsalia of the upper cervical segments and to the topography of the vertebral artery, the posterior inferior cerebellar artery (PICA) and the posterior spinal arteries.

4.3.2 Results

4.3.2.1 Relationships of the SANR to the posterior rootb of the 1st cervical nerve

1. In 100 examined specimens we found macroscopically 73 times a posterior C1 radix.
2. In the 73 specimens that showed a dorsal C1 root there was an anastomosis to the SANR in 46 cases (63%). Thus, in 46% of the normal specimens a sensitive intermixing of the SANR can be assumed. The individual variants of these anastomoses can be divided into 4 types according to OUAKNINE and NATHAN (1973, 1981) (see Fig. 5).

Fig. 5: The different types of anastomosis between the SANR and the posterior root C1



Type I : The posterior root C1 is macroscopically not present (27%).

Type II : The posterior root C1 is present and reaches the medulla oblongata without anastomosis to the SANR (27%).

Type III: The posterior root C1 is anastomotic with the SANR, but continues postanastomotically to its normal entry into the medulla oblongata (34%)

Type IV : After entering the spinal canal, the posterior root C1 anastomoses with the SANR without having a direct connection to the spinal cord.

4.3.2.2 Vascular variations in the cranio-cervical region

In 12 specimens we found an extremely deep outlet of the PICA from the vertebral artery with contact to the SANR. In 7 cases, the PICA already originated extradurally and had its own dural passage dorsal to the dentate ligament. In 5 specimens we found the PICA outlet directly at the dural passage of the vertebral artery, with further course also dorsal to the dentate ligament. In 8 cases the PICA was not located on one side.

A posterior spinal artery was found 65 times immediately after passing through the vertebral artery. In 52 cases the posterior spinal artery ran ventrally, in 4 cases dorsal to the SANR, almost always tangent to it.

4.3.2.3 Summary of the anatomical findings

1. In the craniocervical region, the anatomical variance of the nerve and vascular structures, as well as their topographical relationships to each other is large.

2. Contrary to widespread opinion, a C1 posterior root was present in 3/4 of the normal specimens.

3. In about 45% of the specimens there were neural connections of the posterior root C1 (to a lesser extent also C2) to the SANR. In these cases, the SANR can be described as a mixed nerve. This finding contradicts the view of most anatomical textbooks and the general doctrine (BRODAL, 1957, SOBOTTA-BECHER, 1962, STARCK and FRICK, 1967 and many others).
4. In type IV anastomosis, the afferents of the posterior root C1 reach the CNS via the SANR, whereas this is probable for type III.

5. neurovascular contacts between the SANR and the arteries in the regio craniocervical arteries (vertebral artery, PICA, posterior spinal artery) are common.

4.4 Bilateral microsurgical lysis of the SANRs (BMLA)

Based on the described electromyographic and anatomical findings above, we derived the justification for ST patients with severe, uninfluenceable symptoms to undergo an operation with exposure and inspection of both SANRs. The aim of the procedure was to search for anatomical anomalies that could be the cause of a peripheral genesis of ST in order to eliminate them if possible (FRECKMANN et al., 1981).

4.4.1 Patient population

Since 1980, we have operated on 46 of more than 300 torticollis patients treated on an outpatient basis. All conservative treatment options such as physiotherapy, drug treatment, biofeedback training and in some cases, psychotherapy were exhausted in these patients without any lasting improvement being observed.

9 patients with DT underwent stereotactic surgery. The corresponding treatment results is shown in section 4.5.

In 37 patients where, based on the symptomatology and the EMG findings the presence of a pure ST could be assumed, with the exception of the first three patients, who only underwent unilateral exposure of the accessory nerv, we performed bilateral microsurgical lysis (BMLA) of the SANRs. Findings that torsion dystonia or another extrapyramidal disease were not found preoperatively. There were no anamnestic signs of a possible traumatic, vascular, inflammatory or tumor-related genesis of the disease, with 2 exceptions: One patient had undergone poliomyelitis as a child (case 1), the other had suffered a brain contusion 5 years before (case 6).

The average age of the patients at the onset of the disease was 37.8 years, the duration of the medical history at the time of of the operation was 4.5 years on average. There were 25 men and 12 women.

All 37 patients suffered from a pronounced, often painful ST, in some cases with a tonic fixed head malposition, partly

with involuntary, mostly clonic, but not rhythmic or tremor-like movements of the head to one side. 25 patients showed a predominantly horizontal ST, in 15 patients with turning of the head to the right and in 10 patients to the left. In these patients, the SCM muscle was usually severely hypertrophied on the side opposite to the head turn. Two patients suffered from a purely rotatory torticollis with tilting of the head to the right. 10 patients showed combined torticollis symptoms that indicated involvement of the deep neck muscles: rotatory-horizontal malalignment was found in 6 patients, retrocollis with a horizontal component in 3 patients and anterocollis with a rotatory component in one patient (Table 3).

Of the 10 patients with combined torticollis symptoms, 3 patients also reported severe cervicobrachialgia partly associated with radicular sensory disturbances on the side to which the head rotated. 14 patients were able to influence the torticollis symptoms favorably with the "Geste antagoniste".

All patients underwent computed tomography. CT findings that could have been interpreted as an indication of a central of the disease were not found.

Sporadically performed vestibular function tests revealed no evidence of primary involvement of the vestibular apparatus in the disease.

Vertebral angiography was performed in the first 25 patients. The catheter examination of both vertebral arteries revealed a deep PICA outlet with loop formation towards the caudal side in 11 cases, sometimes reaching below the level of the foramen magnum. No further special features were found. Therefore we have refrained from angiography (FRECKMANN et al., 1986, 1987).

Finally, we conducted a detailed discussion with the patients about the planned procedure and its chances of success and risks. After the appropriate consent was given, the patient was prepared for the operation.

Table 3: Results of BMLA (ST-Patients, n=37)

CASE	SEX	AGE	SYMPTOMS		ILLNESS DURATION (MONTHS)	TYPE OF ANASTOMOSES RIGHT	LEFT	POSTOP-COURSE (MONTHS)	RESULT PERSPECT DOC/PAT.		DT
1	M	37	HOR	L	18	I	-	82	1	1	
2	M	21	HOR	L	18	-	III	63	2	-	
3	M	47	HOR	R	12	III	IV	77	3	3	
4	F	46	HOR	R	48	IV	III	74	2	2	
5	M	52	HOR	L	48	IV	I	46	4	+	
6	M	22	HOR	R	48	II	III	70	2	2	
7	M	36	HOR	L	48	IV	IV	70	2	3	
8	M	26	HOR	L	21	IV	IV	67	1	1	
9	M	42	HOR	L	120	IV	IV	65	1	1	
10	F	25	HOR	R	18	IV	IV	40	3	-	
11	M	41	HOR	L	14	IV	III	57	2	2	
12	M	51	HOR	L	156	IV	III	57	1	1	
13	M	47	HOR	R	18	IV	IV	56	3	4	
14	M	39	HOR	R	30	IV	III	55	3	3	
15	F	38	HOR	R	18	II	III	52	1	1	
16	F	47	HOR	R	42	III	III	46	2	2	
17	M	45	HOR	R	24	III	IV	44	1	1	
18	M	57	HOR	L	84	I	I	33	2	2	
19	M	41	HOR	R	9	IV	III	31	3	3	
20	F	50	HOR	L	18	IV	III	27	3	3	
21	F	24	HOR	R	7	III	III	26	2	2	
22	M	54	HOR	R	78	IV	IV	21	1	1	
23	F	49	ROT	R	36	IV	IV	75	2	3	
24	M	41	ROT	R	108	IV	IV	41	3	+	
25	M	44	HOR	R	144	IV	IV	15	3	3	
26	M	45	HOR	R	264	IV	I	14	2	2	
27	F	45	HOR	R	42	I	I	12	2	3	
28	M	45	ROT/HOR	R	36	I	III	9	3	3	
29	F	48	ROT/HOR	R	120	IV	I	60	4	4	DT
30	F	54	ROT/HOR	L	96	IV	I	49	3	4	
31	M	41	ROT/HOR	L	42	IV	I	46	4	4	
32	M	30	ROT/HOR	L	24	III	IV	40	5	5	DT
33	M	51	ROT/HOR	L	72	IV	III	38	3	4	
34	F	40	ROT/ANT	R	22	IV	IV	42	5	4	DT
35	M	37	RET/HOR	R	36	-	III	82	3	3	DT
36	F	55	RET/HOR	R	42	IV	IV	74	3	4	
37a	M	42	RET/HOR	R	48	III	IV	13	3	4	
37b	M	43	RET/HOR	R	60	-	-	2	+	+	

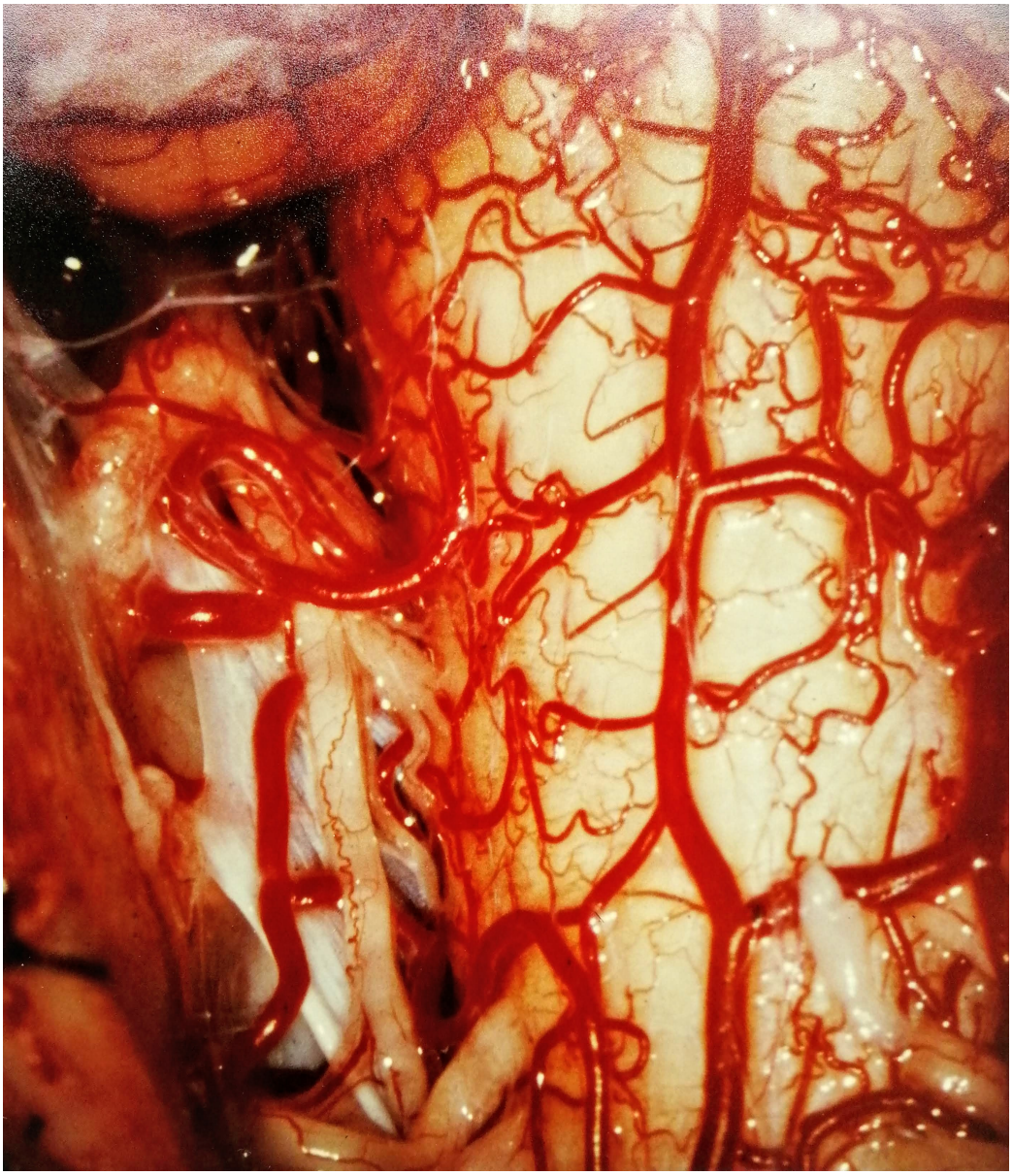
1 (very good) = free of symptoms
2 (good) = painfree, head can hold straight over longer time and is free movable in all directions
3 (improved) = still sometimes unvoluntarily movenments of the head by lack of concentration
4 (unchanged)
5 (worsened) = increase in symptoms or development of Dystonic Torticollis (DT)

4.4.2 Methods

4.4.2.1 Surgical technique

After appropriate preparation, the patients were operated under intubation anesthesia. The procedure was performed in the prone position with fixation of the head in the Mayfield clamp. After a midline incision from the external occipital protuberance to the spinous process of the 2nd cervical vertebra, a small suboccipital craniotomy with laminectomy of the posterior atlas arch was performed. The dura was opened longitudinally so that a precise inspection of the spinal and cranial roots of the accessory nerves down to the level of C2 on both sides was possible. However, the first three patients were only unilaterally exposed. Using the surgical microscope, we searched for microvascular contacts of the accessory roots and for anastomoses between the SANR and the posterior radix of the 1st and 2nd cervical nerves, in accordance with the previous anatomical study. After photo documentation of the anatomical conditions, the anastomoses were cut while preserving the root arteries. In some cases, the posterior C1 radix and, if necessary, the posterior C2 radix were resected and histologically examined, with simultaneous removal of a ganglion often located in the anastomotic area. In 6 cases, during electrical stimulation of the SANRs or the posterior roots C1 and C2, an EMG was recorded intraoperatively from the SCM and trapezial muscles. A micro-nerve stimulator was used for this purpose, as described by MORGAN and JANNETTA (1977) for stimulating cranial nerves in the CPA. After releasing the SANRs from all adhesions and vascular contacts, especially to the vertebral artery, the PICA and the posterior spinal artery, they were protected from further vascular contact by padding with a small piece of teflon foam (Prosthex (R)). The wound was then stopped from bleeding and closed in layers (Fig. 6).

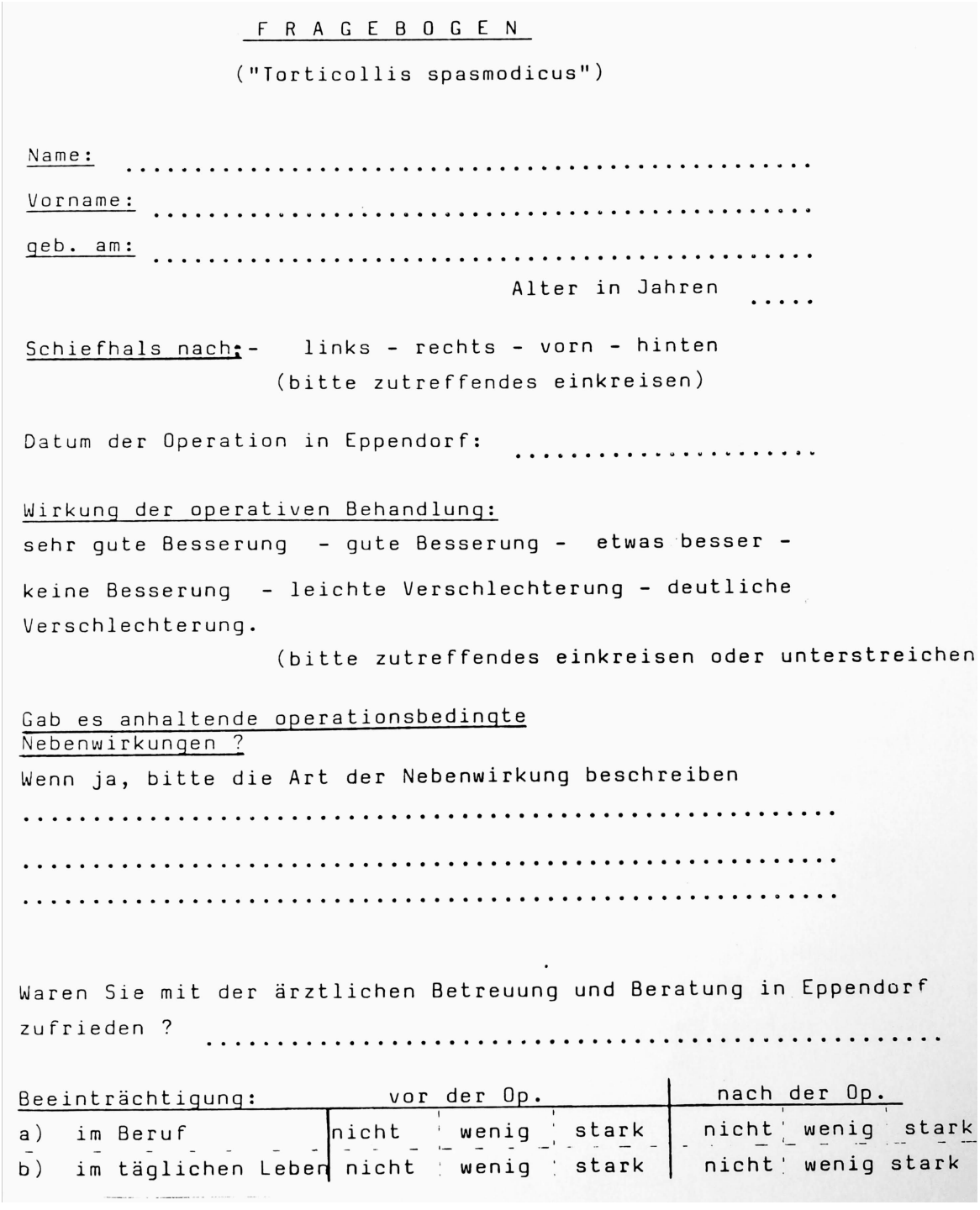
Fig. 6: Case 10, Surgical site at the BMLA, left anastomosis type IV, ganglionous structures at the SANR



3.4.2.2 Postoperative treatment and follow-up

In all cases, the surgical treatment was followed by inpatient, rehabilitative aftercare with special ST gymnastics, exercise baths and adjuvant drug treatment (Muskeltrancopal (R), Dartalan (R), Xylotocan (R)). Outpatient follow-up examinations were carried out regularly 6, 9 and 12 months postoperatively, and thereafter at

approximately annual intervals. In 1982 and 1985, we sent out questionnaires for the ST patients themselves to assess the effect of the surgical procedure (see Fig. 7). At the same time, an outpatient follow-up examination was carried out (HAGNAH et al., 1983, FRECKMANN et al, 1986, 1987)



F R A G E B O G E N

("Torticollis spasmodicus")

Name: .....

Vorname: .....

geb. am: .....

Alter in Jahren .....

Schiefhals nach: - links - rechts - vorn - hinten

(bitte zutreffendes einkreisen)

Datum der Operation in Eppendorf: .....

Wirkung der operativen Behandlung:

sehr gute Besserung - gute Besserung - etwas besser - keine Besserung - leichte Verschlechterung - deutliche Verschlechterung.

(bitte zutreffendes einkreisen oder unterstreichen

Gab es anhaltende operationsbedingte Nebenwirkungen ?

Wenn ja, bitte die Art der Nebenwirkung beschreiben

.....

.....

.....

Waren Sie mit der ärztlichen Betreuung und Beratung in Eppendorf zufrieden ? .....

Beeinträchtigung:	vor der Op.			nach der Op.		
a) im Beruf	nicht	wenig	stark	nicht	wenig	stark
b) im täglichen Leben	nicht	wenig	stark	nicht	wenig	stark

Fig. 7: Questionaire from the follow-up study

4.4.3 Results

4.4.3.1 Anatomical finding

In 1986, we undertook a comparative anatomical study of the topographical relationships between the SANR and the posterior radix of the 1st cervical nerve in ST patients and normal section cases without neurological disease (FRECKMANN and HAGENAH, 1986). This study was based on 71 intraoperative findings obtained in ST patients and the 100 dissection findings described above (see section 4.3).

Analogous to the anatomical findings obtained from cadavers, we found in our ST patients a wide variety of anatomical conditions in the craniocervical region. The relationships between the SANR and the posterior radix of the 1st cervical nerve in ST patients (71 inspected sides) and in the section cases (100 inspected sides) were assigned to the 4 anastomosis types according to OUAKNINE and NATHAN (1973), although different subvariations were observed (see figs. 5., 6., 8., 9., 10.):

Type I : On 11 of 71 sides inspected in ST patients no posterior root C1 was present (15.5%).

Type II : In 2 ST patients a posterior C1 radix was found, each on one side, which without any recognizable anastomosis to the SANR was entering the lateral posterior sulcus (3%).

Type III: An anastomotic connection between the otherwise normal posterior root C1 and the SANR was recognizable on 20 inspected sides (28%).

Type IV : On 38 sides the posterior root C1 entered the spinal canal next to the vertebral artery, connected anastomotically with the SANR and ended at this point, without connection to the lateral posterior sulcus(53%).

We saw equal sides findings in 15 cases. The comparison between surgical and dissection findings is shown in Table 4.

Table 4: Relationships between the SANR and posterior root C1

	DISSECTION CASES	ST PATIENTS
	n=100	n=71
TYPE I	27	11 (15.5%)
TYPE II	27	2 (3 %)
TYPE III	34	20 (28 %)
TYPE IV	12	38 (53.5%)

In addition, we often found a ganglion in the anastomosis area. Sometimes the ganglion was located at the SANR, more often we found it on the posterior radix of the 1st cervical nerve directly lateral to the SANR. In type IV, the SANR often was drawn into the intervertebral foramen C1 and firmly fused with the vertebral artery (see Fig. 6, 9).

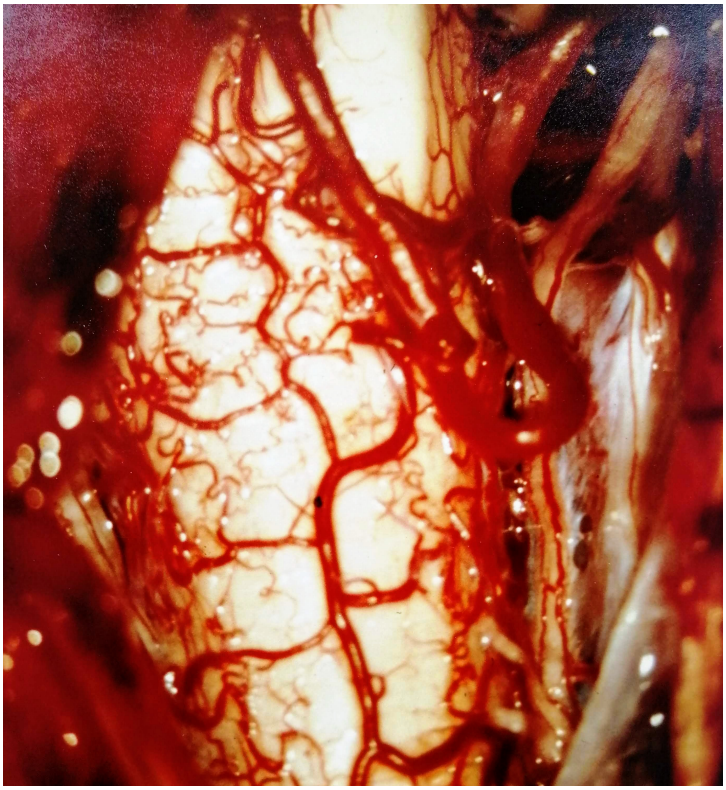


Fig. 8: Case 31, Type I, contact with vertebral artery and deep PICA loop

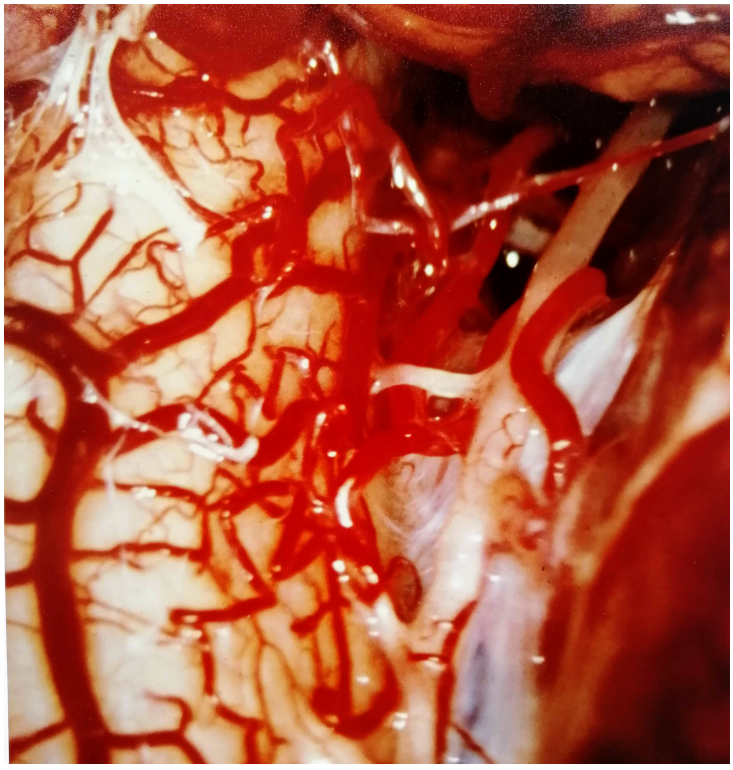


Fig. 9: Case 14, Type IV, ganlion, contact with spinal artery

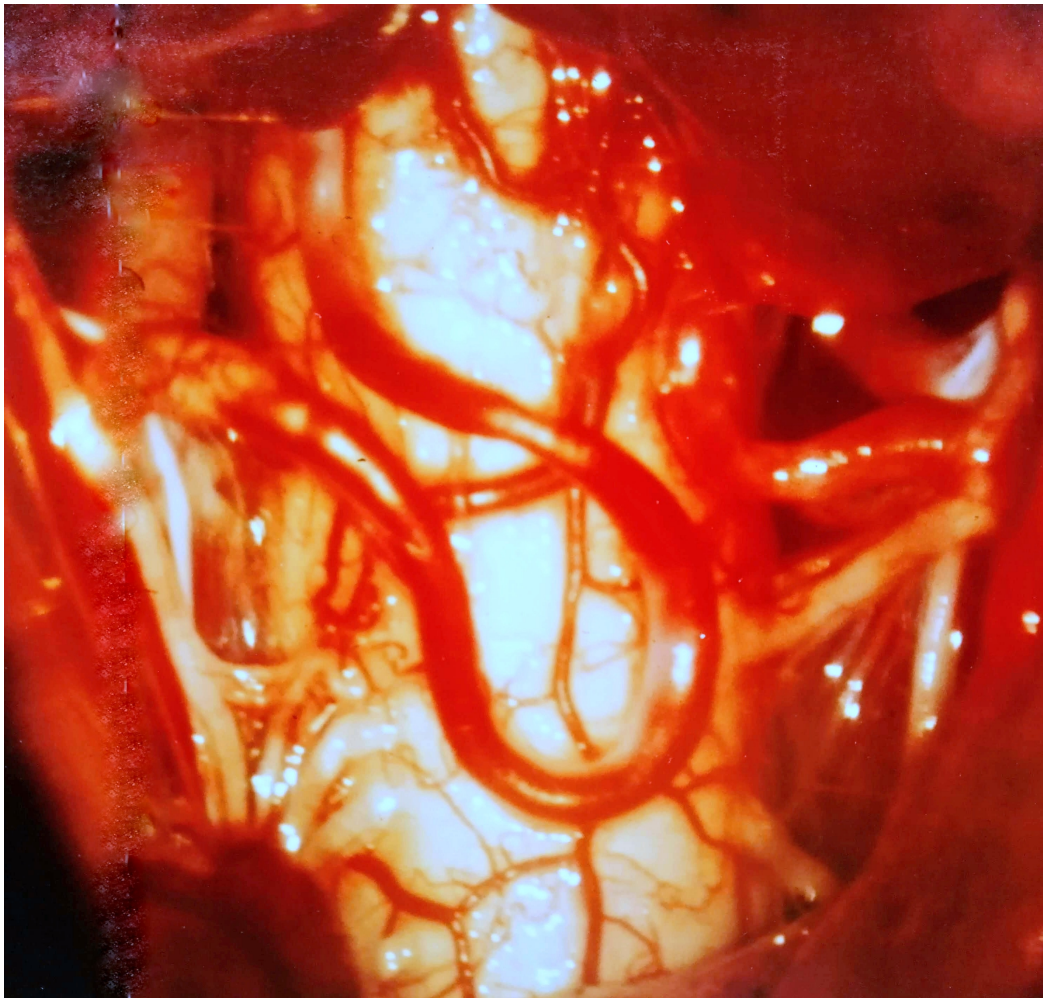
While we found anastomoses between the SANR and posterior root C1 in 46% of the dissection cases, corresponding findings were found in 81.5% of the inspectet sides in ST patients.

In addition, multiple neurovascular contacts were found between the SANR and the arteries of the craniocervical junction, especially to the vertebral artery, to the PICA or to various spinal arteries, in most cases with significant compression of the SANR. In many cases, compression of the SANR by the uppermost dentate ligament was also observed due to the tethering to the vertebral artery. We found vascular contacts of the SANR more frequently in ST patients than in the sectioned specimen. However, the difference is not exactly comparable, as vessels in cadavers do not have any tonus (Tab. 5.).

Table 5: Relationships between the SANR and neighboring arteries in the craniocervical junction

	SECTION CASES	ST PATIENTS
	n=100	n=51
deep PICA outlet	12%	18%
deep PICA loop	0%	20%
Vertebral artery	18%	30%

Fig. 10: Case 3, left type IV and deep PICA sling with with compression of the accessory nerve, right type III



4.4.3.2 Histological findings

The histological examination of the resected posterior roots C1, which maintained an anastomosis to the SANR corresponding to type IV, revealed scarring changes in close proximity to ganglion cell clusters, which were to be regarded as spinal ganglia. In some cases arachnoid villi were found, 4 times we found a small neurinoma or clear Schwann-cell proliferations. In 5 of 7 specimens, signs of degeneration and regeneration were recognizable. These findings were indicative of chronic irritation of the 1st posterior cervical root. In situ, these patients showed firm adhesions of the SANR with the vertebral artery (FRECKMANN et al., 1986).

4.4.3.3 Results of intraoperative nerve stimulation

When we stimulated the SANRs during the first the first 10 operations, we obtained motor responses in the SCM and trapezial muscles. The contraction of both muscles occurred when the accessory nerve root was stimulated above the C2 level. We observed a contraction of the SCM muscle alone when the parts of the accessory nerve root originating at level C1 were stimulated. In contrast, we observed sole contraction of the upper part of the trapezius when the SANR was stimulated below the level of C2. These findings indicate the projection of these muscles in the spinal nuclei.

When the SANR, which is anastomotically connected to posterior root C1 was stimulated it also caused contraction of the SCM muscle in some cases. This finding suggests either the existence of motor parts in the posterior root C1 or for an ephaptic transmission of the impulses to the SANR.

4.4.3.4 Summary of the surgical findings

1. In ST patients, a posterior C1 radix is found in 85% of cases.

2. Anastomoses between the SANR and the posterior root C1, normally present in almost 50% of cases, exist in ST patients in over 80% of cases. In the vast majority of cases, the SANR must therefore be regarded as a mixed nerve.

3. Neurovascular contacts between the SANR and the arteries of the craniocervical junction, especially with the vertebral artery and the PICA, were not significantly more frequent in ST patients than in the general section material. However, there was often a clear compression of the SANR in the anastomotic region.

4. Histologically, two thirds of the specimens showed evidence of chronic irritation of the posterior root C1.

5. Intraoperative stimulation experiments showed that the motor anterior horn cells for the SCM muscle are located at the level of C1, those of the upper part of the trapezius muscle below.

4.4.3.5 Clinical results of the BMLA

In 1981, we first published our results obtained with the BMLA (FRECKMANN et al., 1981). After a mean postoperative observation period of 9 months, an improvement of ST symptoms was observed in 10 out of 11 patients. At the same time, the pain complaints had also regressed in all cases. This result was our most important argument for continuing the surgical treatment of ST by BMLA (FRECKMANN et al., 1983, 1986, HAGENAH et al., 1983).

After postoperative follow-up of now up to 7 years (the average postoperative follow-up follow-up period is 4 years), our treatment results can be assessed as follows (for assessment criteria, see legend in Table 6):

3 of our 37 operated patients have died: One patient (case 37) underwent a repeat surgery 13 months postoperatively, as the ST symptoms had improved only slightly and a painful shoulder-arm syndrome had been added. Two weeks after the operation, the patient was already mobilized, developed medullary symptoms with severe respiratory insufficiency, which led to his death (see also Appendix case 37). The other two patients died 3 1/2 and 4 years after the operation, one in status asthmaticus with pre-existing bronchial asthma, the other as a result of intoxication. Of the remaining 34 patients, we received 32 completed questionnaires. Two patients were unknown moved so that only examination findings that were collected one year postoperatively could be used for the evaluation. In both cases, a good regression of the ST symptoms was recognizable.

The evaluation of the questionnaires revealed that in 7 cases a "very good" result had been achieved. A further 7 patients described the treatment result as "good" and 10 patients felt

that their condition had "improved". The operation had no effect on ST symptoms in 6 patients, while 2 patients reported a worsening of their condition. Surgery-related side effects were noted by 4 patients. In all cases these were unilateral weakness of the arm side elevation.

Table 6: Results of BMLA by treatment of ST

	1	2	3	4	5	SE	DEATH
PATIENTS (n=33)	7 (21%)	7 (21%)	10 (30%)	6 (18%)	2 (6%)	4 (12%)	
		24 (72%)		8 (24%)			1 (3%)
CLINICAL (n=37)	7 (19%)	11 (30%)	13 (35%)	3 (8%)	2 (5%)	4 (11%)	1 (3%)
		31 (84%)		5 (14%)			

1 (very good) = free of symptoms
2 (good) = painfree, head can hold straight for longer time and is in all directions free movable
3 (improved) = still sometimes unvoluntarily movenments of the head by lack of concentration
4 (unchanged)
5 (worsened) = increase in symptoms or development of Dystonic Torticollis (DT)
SE = Side effects

The outpatient follow-up examination carried out parallel to the survey, together with the last examination findings of the two patients who were moved and the two patients who died of unknown causes, gave the following picture: We assessed 7 treatment results as "very good", 11 as "good" and 13 as "improved". In 3 patients the symptoms were considered to be "unchanged", in 2 patients we noted a deterioration (see Table 6, 3). 22 patients (4 before the operation) returned to work, 3 patients were still on sick leave 5, 8 and 9 months after the operation and 9 patients were retired.

The comparison between torticollis type and postoperative outcome showed that the best results were achieved with purely horizontal ST (n=25): Here we saw 24 improvements (96%). The two patients with purely rotatory torticollis

symptoms also improved. Results were poor in the 10 patients with combined torticollis symptoms, such as the rotatory-horizontal type and retrocollis. In 4 cases (40%) there was no improvement, according to self-assessment even in 8 cases (80%). In 4 of these patients, the symptoms increased in the postoperative course, so that the presence of DT had to be assumed.

As an undesirable side effect of the operation we found accessory paresis of varying severity in 4 patients: In one patient, the paresis was complete after intraoperative transection of the SANR, in the 3 other patients there were only mild, largely regressed paresis of the lateral arm lateral elevation (trapezius muscle), without recognizable muscle atrophy.

4.5 Results of stereotactic interventions

4.5.1 Period 1972 - 1979 (undifferentiated patient population)

From 1972 to 1979, 24 torticollis patients underwent stereotactic surgery without prior differentiation of the symptoms (Prof. Müller). Retrospectively, 11 patients had dystonia and 13 patients had syndromes limited to the neck muscles.

All 11 patients with DT (9 men, 2 women) underwent long-term surgical improvement, most of them with tolerable side effects. However, it was not always possible to determine with certainty whether the side effects were exclusively surgery-related or caused by progression of the dystonia.

Of the 13 non-dystonic patients, who retrospectively had a pure ST, only 5 (38%) of patients improved by the stereotactic interventions, 8 remained completely unaffected. Deteriorations were not seen.

In relation to the different stereotactic target points (pallidum or thalamus/subthalamus) there was hardly any difference with regard to the side effects of the operation.

The side effect rate, however, increased significantly with double-sided interventions, although cases without any side effects can also be found here (Table 7.).

4.5.2 Period 1980 - 1987 (DT)

Of the 9 stereotactically operated torticollis patients since 1980 (6 men, 3 women), 8 patients could be reliably assigned to the diagnosis "DT". In one case, a definite classification was not possible. In this cases, there were indications of both a disease triggered by the central nervous system and peripheral neurogenic damage in the EMG. At 72 years of age, this was by far the oldest patient at the time of the operation. Perhaps this explains the uncertainty surrounding the diagnosis. He is the only patient in this group who showed an unsatisfactory result, also with regard to side effects. All other patients in this group improved significantly when they underwent double-sided surgery (78%). The only patient who underwent unilateral surgery (case 27), who works full-time as an engineer, still has minor residual symptoms that are well controlled by medication with 2x5 mg Dartal and 2x10 mg Parkinsan.

These results underline the importance and necessity of the correct differential diagnostic classification of the patients and the corresponding differential therapy (MÜLLER, 1983).

Table 7: Stereotactical surgery results of torticollis

CASE	SEX	AGE	SYNDROM	DURATION MONTHS	TARGETPT.	FOLLOW-UP MONTHS	RESULTS	SIDEEFFECTS
Before 1980								
1	W	51	ST	18	1 obs	3	unaffected suicide	Dysarthria
2	W	50	ST	48	3 R, 1 L	48	sign.improv.	SlightDysarthria
3	M	34	DT	36	3 R, 1 L	36	sign.improv.	-
4	W	42	DT	60	1 obs	150	sign.improv.	slightDysarthria
5	M	43	ST	48	1 L	48	unaffected	opticus-lesion
6	M	36	ST	34	1 R	120	temp.improv.	Spastic part part-Paresis
7	M	32	DT	168	1+2 L, 1 R	48	sign.improv.	DriveDisorder
8	M	41	DT	30	1 obs	96	temp.improv.	-
9	W	25	DT	90	1 R	24	moderat impr	-
10	M	50	DT	60	1 obs	60	sign.improv.	Dysarthria and FinemotoricDisor
11	M	31	DT	204	1 L	48	slight impr.	-
12	M	51	ST	54	1 obs	48	unaffected	DriveDisorder
13	M	31	ST	12	1 obs	24	unaffected	-
14	M	20	DT	24	1 obs	108	moderat impr	slightDysarthria Hemispastic L
15	W	50	ST	54	1 L	36	moderat impr	Insomnia
16	M	54	ST	36	1 L, 3 R	48	unaffected	-
17	M	63	DT	60	3 L	24	moderat impr	-
18	M	38	ST	34	3 L	24	unaffected	-
19	M	40	ST	36	2+3 L	48	slight impr	Part-Par.arm R in C6-C8-Syndr
20	M	38	ST	66	2+3 L	108	unaffected	slightDysarthria
21	M	50	DT	120	1 obs	60	very large improvement	slightDysarthria
22	M	39	ST	36	1 R	36	slight improv.	-
23	M	59	DT	132	1 obs, 2 R	36	signif.improv.	-
24	M	32	ST	12	1 R	36	inaffected	-
25	W	46	DT	18	1 obs	72	signif.impr	slightDysarthria
After 1980								
26	M	72	ST? DT?	132	2+3 R	72	Dyston.improv Tc unaffect	sleight Hemispast.L
27	M	41	DT	48	1 R	48	signif.improv.	-
28	M	43	DT	48	1 obs	42	sign.improv.	-
29	M	17	DT	96	2+3 L, 1 R	30	sign.improv.	slightDysarthria
30	M	18	DT	156	3 R, 1 L	16	moderat impr	Aphonie,tempo-rary Dysarthria
31	M	41	DT	120	1 obs	9	sign.improv.	Pro-/Retropul-sion,Dysarthria
32	W	47	DT	24	1 obs	5	improvement	pass.Hemipar.L
33	W	45	DT	66	1 obs,2 L	5	improvement	Hemispast.R small amount

Target poinST: 1 = Pallidotomy
2 = Thalamotomy
3 = Subthalamotomy
Syndrom: ST = Spasmodic Torticollis without symptoms of Dystonie
DT = Dystonic Torticollis
Illness duration : First symptoms befor OP, in multiple OP until last OP
Follow up: Last follow-up since last Op

5.0 Discussion

While the etiology and pathogenesis of HFS have been largely elucidated in recent times, the cause of ST, in which the accsessory nerve is at least involved, is still unclear.

SCOVILLE and BETTIS (1979), who saw similarities between these two clinical pictures and therefore grouped them under the term "motor tics of the head and neck", made the following assumption: "It is conceivable that the torticollis spasmodicus, like hemifacial spasm, is the result of preexisting vascular abnormalities."

In view of our own unsatisfactory results with stereotactic treatment of clinically undifferentiated torticollis patients, we took up this idea and questiond of whether ST, in contrast to torticolis dystonicus, could also be based on a peripheral neurogenic disorder.

Contrary to the widespread opinion that ST is a central motor disorder, on the basis of our own clinical, anatomical, electroneurophysiological and surgical findings, we came to the conclusion, divided into several points, which I am putting forward for discussion as an alternative theory on the pathogenesis of ST:

1. Clinically, a distinction must be made between "pure" ST and DT, which belongs to the group of torsion dystonia. These are two different, etiologically separate clinical picture. The EMG reveals different innervation patterns for ST and DT. It therefore facilitates the differential diagnosis.

2. All neuropathological findings obtained to date and the treatment results achieved to date speak, different than in DT, against a central motor genesis of ST. The cause of ST therefore has to be peripheral, in the area of the motor terminal pathway and/or the proprioceptive afferents responsible for head motor function.

3. The following findings support a peripheral genesis of ST:

a) The electromyographic derivation of individual motor units from the muscles supplied by the accessory nerve revealed reproducible abnormal single potentials, which can be interpreted as evidence of a peripheral neurogenic lesion.

b) The posterior root C1 macroscopically is normaly found in 73%, in patients with ST in 85% of cases.

c) In ST patients are found far more frequently than in the normal collective (82% : 46%) anastomotic connections between the SANR and the posterior root C1. Thus, the accessory nerve is a mixed nerve in these cases.

d) The sensory parts of the accessory nerve are proprioceptive afferents from the head joints and from the muscles which are supplied by the accessory nerve.

e) In ST patients, intraoperatively in 2/3 of the cases there were found significant compression of the SANR with evidence of degenerative processes, which were confirmed histologically.

The latter findings indicate an anatomical predisposition in ST and allow the conclusion of a peripheral genesis.

4. Accordingly, the surgical treatment of ST requires a new peripheral approach that differs from TD:

The BLMA is aimed at the elimination of unilaterally disturbed craniocervical, proprioceptive afferents in order to symmetrize the impulses to the reticular formation and thus reduce thereby influencing the ST symptoms. The procedure is less destructive and has hardly any side effects.

5. The results of the surgical treatment of torticollis can be significantly improved if

a) a clinical differentiation is made between ST and DT

and if

b) both clinical pictures are treated differently according to their aetiology: ST with the peripherally applied BMLA, the DT with centrally performed stereotactic surgery.

Before I present the arguments in favor of a peripheral genesis of ST in detail, the pathogenesis hypotheses that have been discussed so far in ST will be described briefly and the reasons for their untenability are given.

I will refrain from discussing a psychological cause of ST, since today, with a few exceptions, most authors agree on an organic genesis of the disease (see p. 20).

The hypothesis of a labyrinthine or vestibular genesis of of ST is based on the observation that in some patients unilateral pathologic vestibular or labyrinth functions, in the form of nystagmus opposite to the direction of the ST are present. To date, however it has not been clarified whether this is a primary or secondary involvement of the vestibular system. Overall, a vestibular genesis of ST must be regarded as unproven and, apart from a few exceptions, as unlikely (see p. 21).

The formatio reticularis hypothesis is based on the idea that small, only quantitatively measurable changes in the area of the reticular formation lead to an interruption of inhibitory influences on the equilateral vestibular nucleus and thus to the development of ST symptoms. A pathologic-anatomical substrate for this hypothesis has not yet been found. There is no doubt that the reticular formation plays an essential role in the coordination of head motor function and the senses of balance. There is nothing to support the assumption that the lesion responsible for ST is localized in the formatio reticularis itself (see p. 35).

The basal ganglia hypothesis, first formulated by FOERSTER in 1920, has found the broadest support of all pathogenesis

hypotheses. Almost all recent publications begin with the statement that ST is probably the focal form of a central motor movement disorder, although the underlying striatal damage of these disorders underlying striatal damage in ST could not be proven. Pathological findings in the CNS were only described in cases where, apart from the symptom "torticollis", clear indications of an extrapyramidal dystonic movement disorder (DT) were present. In the cases where, according to the casuistry, a pure ST is to be assumed, there were no pathological findings in the area of the basal ganglia (see p. 24). CT also shows normal intracranial conditions in the ST patients.

Apart from the difficulty of performing animal experiments specifically conducted to produce a head malposition, transferring to the human ST, stereotactic lesions in the basal ganglia are more likely to serve as a model for post ischaemic athetosis (see p. 34).

Drug treatment trials with cholinergics and dopaminergics, which are analogous to the treatment of Parkinson's disease on the neurotransmitter system of the diencephalon and the mensencephalon, also speak due to their proven ineffectiveness against a primary involvement of the basal ganglia in the pathogenesis of ST (see p. 57).

The unsatisfactory results achieved in ST with stereotactic interventions also speak against the basal ganglia hypothesis (see p. 48).

In my opinion, the basal ganglia hypothesis has to be abandoned today, almost 70 years after the postulate of striatal damage in the genesis of ST and after many unsuccessful attempts to prove it, must be regarded as untenable. In order to do justice to the problem "ST", the entire regulatory circuit responsible for the control of head motor function must be considered. This includes not only the central structures such as the basal ganglia and the reticular formation, but but also the sensory organs, the muscle efferents and afferents, the motor units and the

proprioceptors of the cervical and head joints.

In conclusion, all previous efforts to investigate the cause of ST have failed. ZEMANN and WHITLOCK therefore suspected as early as 1968 that that "pure" ST may have a different etiology, since its treatment, in contrast to torsion dystonia had proved to be particularly problematic.

Differential diagnosis of ST

The boundary between ST and torsion dystonia is often not clearly drawn in the literature. We too did not made a precise distinction between the two clinical pictures until 1980. Often all patients with torticollis symptoms were indiscriminately grouped together under the diagnosis of ST. This may be one of the reasons for the overall unsatisfactory treatment results in ST (KRAYENBÜHL u. YASARGIL, 1965, COUCH, 1976 BERTRAND and MOLINA-NEGRO, 1986). MUNDINGER and coworkers, for example, never listed the pure ST and the DT separately in their publications (KRAYENBÜHL and YASARGIL, 1965).

The importance of careful clinical differentiation of ST for the therapy was already highlighted by CASSIERER, 1922, OLIVECRONA, 1931 and KAPPIS, 1934, later also by ARSENI and MARETSIS, 1971, OJEMANN and WARD, 1973, NITTNER, 1986, SHIMA et al., 1986, 1987 and VAN HOOF et al., 1987. LÜCKING (1980), who described ST unchanged as a focal form of dystonia, still found, on the basis of of electromyographic findings that this be regarded as an independent clinical picture within the extrapyramidal motor disturbances.

ST is defined as a movement disorder limited exclusively to the neck muscles (see p. 7). In all torticollis patients must therefore be searched for additional symptoms. Torticollis in conjunction with oromandibular dyskinesia, blepharospasm or signs of tremor in the extremities is highly suspicious for the presence of DT.

In an undifferentiated patient population of torticollis

patients, additional movement disorders were found in 10-20% of cases, indicating the presence of torsion dystonia (KRAYENBÜHL and YASARGIL, 1965, COLBASSANI and WOOD, 1986).

A further indication that can be used for differential diagnosis gave FRIEDMANN and FAHN (1986), who examined 450 patients with torsion dystonia of any type with lasting remission in the order of 12%, only in those cases, what they called "focal" dystonia, which they regarded as “idiopathic".

However, one must ask oneself whether ST is not possibly the precursor of dystonia. After all, BRÄUTIGAM (1954) found that in 20% of his cases with the leading symptom "torticollis", the symptoms later progressed to torsion dystonia. In this respect he spoke of a transitory ST. BERTRAND (1987) also observed 9 patients among 111 "pure" ST cases in which "adult onset dystonia" developed from "classic" rotatory torticollis within a few years. Based on this experience, surgery should not be performed too early. The development of dystonia, which has a progressive course, is different from that of ST, which can be considered complete after 2 years (BERTRAND, 1987). In some cases, however, the diagnosis of "ST", even with the greatest care, is not possible with absolute certainty, misjudgements occur again and again.

In our patient population we observed a postoperative progression of symptoms in 4 of 37 patients to torsion dystonia. Interestingly, these were exclusively patients with combined, clonic torticollis symptoms. Today we are therefore of the opinion that a primary combined torticollis symptomatology, even if no other symptoms not yet recognizable, can be an expression of an incipient DT (FRECKMANN et al., 1986, 1987).

These observations alone underline the necessity, but also the occasional difficulty, to separate ST from DT clinically, which, however is of fundamental importance for adequate surgical treatment.

The clinical analysis of the two clinical images is supported

by EMG, as the ST and DT have different innervation patterns, which facilitates the differential diagnosis. In addition to the clinical examination, an EMG should be performed in each case. Whereas in DT, as in torsion dystonia, both agonistic and antagonistic muscles and muscle groups are involved in the symptoms, in pure ST, only agonists are involved in the symptomatology. However, both sides are practically always affected in both diseases. In ST, the most common is the interaction of the ipsilateral SCM muscle and the contralateral trapezius and splenius, which results in a purely horizontal ST (HERZ and HOEFER, 1945, PODIVINSKY, 1968, FASSHAUER, 1980, LÜCKING, 1980, BERTRAND et al., 1978, 1982, BERTRAND, 1987, HAGENAH et al, 1980, 1981, 1983).

The neurogenic ST

All previous clinical and neuropathologic findings (see p. 25) and the treatment results achieved (see p. 63), in contrast to DT, speak against a central motor genesis of ST.

In terms of its appearance, ST is more similar to the symptomatic torticollis forms than to DT. In tumor diseases located in the foramen magnum area, symptoms have been described that are completely similar to those of ST (KRAYENBÜHL, 1973). In these cases the cause of the torticollis symptoms was either an irritation of the caudal brainstem or the nerve structures in this region, especially the SANR in this region. The case described by FOERSTER (1929), in which a vertebral arterie aneurysm with contact to the SANR led to typical ST symptoms. According to observations by PATTERSON and LITTLE (1943) and DENNY-BROWN (1962), torticollis symptoms in connection with vascular dysplasia are not rare.

The symptomatic forms of torticollis, in which tumors or vascular dysplasia have been identified as the cause of torticollis symptoms therefore suggest a peripheral cause in the area of the motor terminal pathway and/or the proprioceptive afferents responsible for the head motor function localized noxious agent in the pathogenesis of ST.

A predominantly peripheral cause of ST has hardly been considered so far.

Findings in favor of a peripheral genesis of ST

As the accessory nerve is a cranial nerve, and since there are certain similarities between ST and HFS, 1980 we started with our torticollis patients, hypothesizing a mechanical irritation of the SANRs, to look for evidence of a peripheral lesion of the nerve, which, as in HFS, could be a possible cause of the ST. EMG, as a non-invasive method, seemed particularly suitable.

In fact, after modifying the electromyographic recording technique with the aid of bipolar needle electrodes, as required to assess the potential size, width and shape of individual motor units (see p. 67), in over two thirds of our torticollis patients reproducibly pathologically altered potentials from the SCM and trapezius muscles we were able to deduce. Thereby discrete findings compared to the usual EMG derivations were classified as suspicious, since in the case of such irritations only minor neurogenic damage can be expected in such irritations anyway. These findings, which were considered to be indicative of an accessory nerve lesion, support the notion of a peripheral neurogenic factor in the pathogenesis of ST (FRECKMANN et al., 1981, HAGENAH et al., 1981, 1983).

SHIMA et al. (1987) also found EMG changes in one of 4 ST patients, suggesting a neurogenic lesion of the SCM muscle.

As the EMG findings in ST from the muscles supplied by the accessory nerve do not reliably prove neurogenic damage, today we are very cautious in the interpretation of these EMG findings (FRECKMANN et al., 1986, 1987).

In our anatomical studies, which, with knowledge of the suspicious EMG findings, also aimed at finding the causes for a possibly mechanical irritation of the SANRs, in addition to a wide variety of vascular courses, often with close contact

to the SANR, especially the manifold anastomotic connections of the SANR with the posterior roots C1 and C2 impressed. These lead to the conclusion that in the vast majority of cases the SANR is a mixed nerve. Remarkable are also the ganglionic structures associated with the SANR. In addition, we often found nerval onnections of the posterior roots C2 and C3, to a lesser extent also between C1 and C2, which are referred to as intersegmental anastomoses (KOSAK et al., 1981, HAGENAH et al., 1983, FRECKMANN and HAGENAH, 1986) (see p. 71, 80).

KAZZANDER (1891) was one of the first to investigate the question of whether the accessory nerve was purely motor or of mixed character and whether its roots contain ganglionic structures. He found anastomoses between the dorsal C1 root and the SANR in 32% of the specimens, in a few cases also to the posterior root of the 2nd cervical nerve. He concluded that in these cases there was a reciprocal exchange of nerve fibers between the posterior root of the first cervical nerve and the accessory nerve. From the fact that in individual cases the posterior root of the first cervical nerve even originates directly from the accessory nerve, it can be concluded with regard to the functional character of the accessory nerve that it is not exclusively motor, but also originally contained sensory fibers. These findings were later confirmed by PALLIE (1959), OUAKNINE and NATHAN (1968) and STRELKA (1978), who found anastomoses between the SANR and the posterior root C1 in 50% of a normal section good.

A classification of the different manifestations of anastomotic connections of the accessory nerve with the posterior posterior roots of the proximal cervical nerves was first made by (WEIGNER (1901). He also described prominent swellings at the connection points of the SANR to the posterior root C1 which, histologically, represent a sheathing of the nerves with fibrillar connective tissue, but in the nerve trunk itself with spindle-shaped ganglion cells. WEIGNER came to the conclusion: "The posterior root of the 1st cervical nerve can be replaced by root fibers of the spinal accessory nerve. In the spinal and also in the bulbar

accessory nerve roots ganglion cells occur especially when the 1st cervical posterior root is substituted by the accessory nerve. These are not only microscopically recognizable ganglion cell groups, but also macroscopically recognizable ganglia. Herefore, macroscopically the intervertebral ganglion primum is very often absent. In addition, various intersegmental anastomoses between the dorsal roots of the cervical nerves were found."

As part of a study on the embryonic development of the brain and spinal nerves, STREETER (1905) and later also PEARSON (1937) found a close relationship between the SANR, which also had bundles of ganglion cells, to the ganglion of the 1st cervical nerve. In early development, the vagus nerve and the accessory nerve develop from a complex in which both parts had both motor and sensory elements. Only in later development, the vagus becomes a predominantly sensory nerve and the accessory a predominantly motor nerve. Anastomoses between the posterior root of the 1st cervical nerve and the SANR, which can undoubtedly be explained by embryonic development, were found by STREETER in 62% of the cases.

With degeneration experiments on rats and mice, FITZGERALD et al. (1982) investigated the question of the sources from which comes the innervation of the neuromuscular spindles in the SCM and trapezial muscles. In comparison to all other somatotropic skeletal muscles, the SCM and trapecius muscles are unusual in terms of their motor and sensory supply. This because they are supplied via 2 pathways: The entire motor supply of both muscles, both extrafusal and intrafusal, takes place via the accessory nerve. The sensory supply of the corresponding muscle spindles, on the other hand, takes place via the upper cervical nerves and the caudal part of the trapezius via the upper thoracic nerves.

CORBIN and HARRISON (1938) were able to show in a similar way in cats with stimulation- and degeneration experiments that the sensory parts of the SANR and the upper posterior cervical roots are predominantly proprioceptive elements. They came to the the conclusion that afferent fibers from the

superior cervical ganglia both directly and after association with terminal branches of the accessory nerve, go into the muscles supplied by the accessory nerve and supply the corresponding muscle spindles. In the section of the jugular nerve directly outside the jugular foramen, they found no sensory nerve components. In cats therefore, the addition of proprioreceptive fibers to the accessory nerve occurs exclusively extraspinal (CORBIN et al., 1936, HINSEY and CORBIN, 1934). However, this view is not correct. WINDLE (1931) was able to show that the SANR is a mixed nerve in both, in the monkey and the cat. It is correct, however, that the accessory nerve directly after its cranial exit is of predominantly motor quality, since most of the afferents, as shown, the accessory nerve only intraspinal reach the anastomosis with the upper cervical posterior roots.

For humans, VILLIGER (1964) and LANG (1981, 1982, 1983) confirmed the proprioceptive supply of the SCM and trapezial muscles via the 1st, 2nd and 3rd cervical nerves. They determined that the accessory nerve is a mixed nerve. The accessory nerve carries predominantly motor, but also undoubtedly sensory fibers for proprioceptive supply of the upper cervical joints and the muscle spindles of the muscles supplied by the accessory nerve.

RANSON et al. (1932) and CORBIN and HINSEY (1935) found in cats that the afferents of the posterior root C1, to a lesser extent C2 and C3, enter the vestibulospinal tract. In contrast, the posterior root C4 plays no role in the maintenance of the tonic cervical and flexes. Similar findings were reported by MYSICKA and ZENKER (1981) in rats. They found the endings of the muscle afferents from the SCM muscle in the medial parts of the posterior and anterior horns at level C1 to C3, as well as in the reticular formation, indicating a close relationship of proprioceptive afferents from the craniocervical region to the afferents from the vestibular nuclei.

Also interesting in this context are experiments by HÜLSE (1983) as part of a study on cervical balance disorders.

With unilateral transection of the posterior roots C1 and C2 in rabbits, he was able to demonstrate a rotation of the head towards the operated side. This finding was contrasted with an experiment by COHEN, who, after bilateral transection of the posterior roots C1-C3 in monkeys, no lateral deviation found. This only showed a restriction of dexterity in climbing behavior.

I therefore agree with SVIEN and CODY (1969) insofar as it is possible to explain the symptom "torticollis" by reducing afferent impulses to the reticular formation. The reported successes with iontophoresis of the middle ear for the treatment of ST therefore also support our view that the causative disorder in ST must be localized predominantly in the peripheral, afferent leg of the motor function of the head.

Also of importance in this context are clinical observations of patients with tumors in the area of the foramen magnum who exhibited torticollis symptoms and complained of cervico-occipital pain (KRAYENBÜHL, 1973). Since these tumors did not extend down to the level of the 2nd cervical root., the pain could not be be explained by compression of the 2nd cervical root. After discussing the problem with KUBIK, KRAYENBÜHL came to the following explanation:
The anterior and posterior roots of the first cervical nerve are already connected intradurally, so that the nerve only has one dural passage. Extraspinal the suboccipital nerve innervates the short suboccipital muscles and gives off a branch to the atlanto-occipital joint. The posterior ramus of this nerve is often erroneously described as purely motor. The existence of this joint branch proves the opposite. The neck pain in these patients can therefore be explained by an irritation of the posterior root C1.
As the clinic shows, the posterior root of the 1st cervical nerve does not normally provide a sensory supply to areas of skin. ROHR (1963), however, was able to demonstrate in one of one of six rhizotomized ST patients a circular loss of sensitivity at the back of the head, which he attributed to the severing of the 1st cervical posterior root. OBERLÄNDER

(1975) also found during the dissection of a cadaver a sensory skin branch from the dorsal root of the 1st cervical nerve to supply the back of the head.

In ST patients who had undergone a cervical rhizotomy Mc KENZIE (1955) was the first to describe the anastomoses of the SANR to the posterior root C1. He found them in 5 out of 10 cases. KRAYENBÜHL and YASARGIL (1965) emphasized in their discussion of FOERSTER/DANDY's operation that these anastomoses must always be severed in order not to jeopardize the success of the operation. This could explain some of the failures of cervical rhizotomy for the treatment of ST (PALLIER, 1959, OUAKNINE and NATHAN, 1968, STRELKA, 1978).

It is also interesting to note HAYWARD's (1986) observation that in 9 of 15 ST patients, after performing a bilateral rhizotomy of the anterior roots C1-C3 with transection of the SANRs, contractions in the SCM muscle were still present, so that further operations were necessary. This finding suggests that in ST patients there may be a deviation from the normal innervation of the muscles responsible for head rotation.

Some publications deal specifically with the the clinical significance of the intersegmental anastomoses between the posterior roots of the spinal nerves: The existence of such connections can be explained by embryological factors, such as the existence of interganglionic bridges between the spinal nerves and the functional relationship between the upper cervical nerves and the accessory nerve (SCHWARZ, 1956, LANG, 1981). These anastomoses are also the reason for some deviations from the "normal" dermatome scheme as far as the distribution of sensory deficits and pain sensation is concerned (SCHWARTZ, 1956, PERNECZKY and SUNDER-PLASSMANN, 1980). The intraoperative identification of these anastomoses, which occur most frequently in the lower cervical region and in the lumbar region, is particularly important for the success of dorsal rhizotomies for pain treatment (PALLIE, 1959).

COGGESHALL (1979), on the other hand, made an obviously not

100% separation of afferent and efferent nerve fibers for the failures in the treatment of uninfluenced pain, since he used histological methods to identify a large number of unmyelinated fibers in the radix spinalis anterior.

On the course and variations of the vertebral artery, the PICA and the posterior spinal artery can be found detailed descriptions in the literature (GREITZ and SJÖGREN, 1963, LANG and v.WACHSMUTH, 1979, LASJAUNIAS and MANELSE, 1979, FU-JII et al., 1980, LISTER et al., 1982, LANG, 1981, 1985). Specifically on the question of vascular contacts between the SANR and the arteries in the area of the craniocerebral junction there are no studies available.

The quantitative comparison of accessory-vascular contacts, which we found in a normal dissection specimen and in our ST patients, contrary to initial assumptions no significant differences were found (HAGENAH et al., 1983). Nevertheless, as the cause of ST a mechanical irritation of the SANR must be assumed, especially if there is an anastomotic connection with the posterior root C1. In these cases, which occur with a frequency of 82% in ST, the SANR is located at the level of the atlanto-occipital membrane, often even pulled into the C1 foramen and firmly adherent there.

Taking into account the complicated biomechanics in this very mobile region, as investigated by BREIG (1964) and BREIG et al. EL-NADI (1966), a mechanical irritation of the SANR at the level of the occipital foramen magnum is to assume, because considerable torsion, distraction, shear and compression forces occur here.

In fact, we intraoperatively in many cases found a macroscopically visible compression of the SANR. This was stretched either via the vertebral artery, a deeply arising PICA or via the uppermost attachment of the dentate ligament. The SANR was flattened and sometimes, especially when ganglionic structures were attached to it, strongly vascularized (FRECKMANN et al., 1986, 1987) (see p. 82).

In the histologic examination of some C1 posterior roots, which showed an anastomosis to the SANR corresponding to type IV, in 2/3 of the specimens we found clear indications of de- and regeneration processes, which were interpreted as an expression of chronic irritation. In addition, we found fibrous changes in close proximity to ganglion cell clusters, as already described above. In 4 cases we found clear Schwann-cell proliferations, which looked like a small neurinom (FRECKMANN et al., 1986, 1987).

The following finding from the literature is worth mentioning in this context (FINNEY and HUGHSON, 1925: quote BYRNES): In a ST patient, the SANRs had been resected. The histologic examination revealed swelling and degenerative changes in the axons and total demyelination of the nerve. Accordingly degenerative changes were also found in the SCM muscle. The vascular and ligamentous compressions of the SANR that we found intraoperatively in our ST patients differ significantly from those we found in patients with HFS and TN, at the root exit and entry zone of the facial nerve and the trigeminal nerve. Compression of the SANR by the vertebral artery or the upper dentate ligament attachment is to a much greater extent than microvascular compression, as we know it with cranial nerves in the CPA.

Such nerve compressions are considered to be the cause of focal demyelination. These in turn can lead to interactions between neighboring, myelinated axons, which are referred to ectopic impulse generation or ephapses (UEXKÜLL, 1894, GRANIT et al., 1944, MARRAZI and LORENTE, 1944, FRANKENHÄUSER and NYSTRÖM, 1954, GARDNER, 1962, GARDNER and SAVA, 1962, RYDEVIK and NORDBORG, 1980).

Taking into account our surgical and histologic findings, I consider a focal demyelination of the SANR, especially its proprioceptive parts, with the development of an ephapse, as a causative factor in the genesis of of ST.

This view is also supported by experiments in rabbits made by RYDEVIK and NORDBORG (1980), who induced a tibial nerve

compression by a dosed compression of the tibial nerve to produce focal demyelination. Similar experiments were also carried out by BURCHIEL (1980, 1981) conducted on the trigeminal nerve root of monkeys, where he demonstrated abnormal impulse generation after focal demyelination.

Summary of the anatomical findings

The accessory nerve, the 1st, but also the 2nd and 3rd cervical nerves, are part of a closely linked functional system for the control of head motor function. The afferents of the 1st, less also of the 2nd and 3rd cervical nerves propriozeptively supply the head joints, the tension receptors of the SCM muscle and the cranial part of the trapezius. They thus represent the sensory "counterpart" of the accessory nerve (FITZGERALD et al., 1982). This is supported by anastomoses between the SANR and the posterior roots of of C1 and C2. In cases where macroscopically visible no C1 posterior root is present, its function is probably taken over by the accessory nerve and by the C2 root (OUAKNINE and NATHAN, 1973, LANG and von WACHSMUTH, 1979, LANG, 1881).

These anatomical variations also speak in favor of the by OLIVECRONA (1931) PALLIE (1959), KRAYENBÜHL and YASARGIL (1965), OUAKNINE and NATHAN (1968) and STRELKA (1978) supportet assumption, that the intraspinal anastomoses between the SANR and the posterior roots of the superior cervical nerves, especially the 1st cervical root, are responsible for the failures of the rhizotomy according to FOERSTER/DANDY.

According to our investigations, the posterior root C1 is is usually present in 73%, in ST patients even in 85% of cases. The difference is even clearer with regard to the anastomotic connections between the SANR and posterior root C1. In ST patients, these anastomoses are found almost twice as often than in the normal collective (82% : 46%). This proves that the accessory nerve is a mixed nerve. In the sensory parts of the SANR are proprioceptive afferents from

the head joints and the tension receptors of the muscles supplied by the accessory nerve (LANG, 1982).

The anastomoses to the posterior root C1 require fixation of the SANR at the level of the atlanto occipital membrane. This prevents mechanical impairment of the SANR in the form of strong, localized compression of the nerve by the vertebral artery, a deeply arising PICA, the uppermost attachment of the dentate ligament or also by the bony edge of the occipital foramen magnum. In more than two thirds of our ST cases, one or both SANRs showed clear signs of compression with evidence of degenerative processes. Our histologic examinations of surgically removed C1 posterior roots revealed in most cases the existence of a small ganglion in close relation to the anastomotically connected accessory roots. We also found a small neurinoma or clear Schwann-cell proliferation. 5 of 7 findings indicated de- and regeneration processes and thus spoke for chronic nerve irritation. Focal demyelination in the area of the SANRs or the posterior roots C1 with development of an ephapse, as pathogenetic agent in ST, is probable at least for most of our operated cases.

LANG (1982) assumed that the removal or severing of the proprioceptive afferents, which are mainly transmitted via the upper posterior cervical roots to the ganglionic structures at and in the SANR, could result in a change of the clinical picture. Our operation findings primarily suggest an irritation or lesion of these afferents in the area of the anastomoses between the SANR and the cervical posterior roots. Consequently, the cause of a centrally controlled compensatory malposition of the head is a unilateral disturbance of these afferents from the head joints and from the muscle spindles of the of the SCM and trapecius muscles is conceivable.

However, it is also clear from this, that the anatomical features cannot be the sole cause of the ST. Rather, these findings and considerations, as SCHALTENBRAND (1935) already suspected, point to an anatomical predisposition in ST,

which, for hitherto unexplained cause in midlife from latency leads to ST: "Although it is likely a special constitution necessary for the development of torticollis, but on the other hand the cause of the disorder could also lie in a muscle disease that causes chronic irritation of the proprioceptors and in predisposed individuals triggers the peculiar spasms of the neck muscles."

A new operational approach

In the editorial of the YEARBOOK NEUROLOGY, NEUROSURGERY 1980 the discussion of microvascular decompression, the following assumption is made: "It is possible that the responsible lesion for the genesis of torticollis spasmodicus is caused by an arterial loop that leads to unilateral compression of the medulla"(Interurban Neurosurgical Society,February 1978). Apart from this, the literature, with the exception of of SCHALTENBRAND (1935) already cited, SVIEN and CODY (1969), SCOVILLE and BETTIS (1978) and DIAMOND et al. (1987), no further considerations dealing with a a possible peripheral genesis of the ST are found.

On the basis of all currently available knowledge about ST, as well as knowledge of the anatomical and neurophysiological characteristics of the complex control of head motor function and balance, which indicate an anatomical predisposition in ST, the conclusion of a peripheral genesis of this of this disease is justified in my opinion. As a result, ST requires, different to DT, a new peripheral surgical approach (FRECKMANN et al, 1981).

For other cranial nerve dysfunction syndromes (CDS), such as HFS and TN, JANNETTA (1980), after preparatory work by DANDY (1938) and GARDNER (1962), was able to demonstrate the causal significance of neurovascular contacts at the entry and exit zones of the corresponding cranial nerves. Under the assumption a pathogenesis similar to that of HFS, therefore in patients with massive ST symptoms that could not be influenced by conservative methods, we decided to expose and inspect the cranio-cervical region.

Before introducing our new surgical method, the BMLA, we first developed the hypothesis of a peripheral disturbance responsible for ST, a peripheral disorder of the efferent motor limb of the reflex arc responsible for head motor function. We therefore initially searched, as in HFS, primarily for vascular contacts of the SANR (FRECKMANN et al., 1981). With increasing surgical experience, however, we saw that the normal anatomical findings differed considerably from those in our ST patients. However, this was not so much in relation to the neurovascular contacts, but rather with regard to the anastomotic connection between the SANR and the and the upper posterior cervical roots (FRECKMANN et al., 1983, 1986, 1987). However, we already performed our first operations, with the exception of the first three unilateral procedures, in addition to the vascular decompression of the SANRs, the bilateral transection of these anastomoses was performed (FRECKMANN et al., 1981) (see p. 77).

These surgical findings, which surprised us, led us to the the conviction that ST is less often a disturbance of the efferents, but more often an irritation of the afferents from the proprioceptors of the head joints and from the tension receptors of the muscles supplied by the accessory nerve. The assumption of pathogenic vascular contacts with exclusive irritation of the efferent parts of the accessory nerve could not be fully maintained, even on the basis of the findings of LANG (1985) (FRECKMANN, 1986, 1987).

Based on our results obtained with the BMLA in 37 ST patients, however, we feel confirmed in the view that in the pathogenesis of ST, especially when ST symptoms are purely horizontal, a peripheral triggering factor is present. We therefore now believe that the cause of ST lies predominantly in the afferent part of the head motor system (FRECKMANN et al., 1986, 1987).

Our surgical aim therefore is, in addition to the elimination of mechanical irritations in the area of the SANRs, the bilateral interruption of the proprioceptive stimulus flow from the head joints and the tension receptors of the

musculature supplied by the accessory nerve in order to make the remaining afferent stimulus flow to the reticular formation symetrically.

The delayed postoperative regression of ST symptoms, also described by CURSCHMANN (1907), CASSIERER (1922), FINNEY and HUGHSON (1922), FRAZIER (1930) and SHIMA et al. (1987) seems at first to speak against our theory. However, "false", chronically ingrained movement patterns are conceivable, which can only be slowly corrected by intensive physical training. We therefore emphasize the the need for special physiotherapeutic follow-up over a longer period of time.

Results of surgical treatment

Up until 1980 we had unsatisfactory overall treatment results with the exclusively stereotactic treatment of a mixed patient population consisting of 11 patients with patients with DT and 13 patients with ST (FRECKMANN et al., 1981, MÜLLER, 1983). Out of 24 patients, only 16 (66.6%) could be improved with tolerable side effects. The results with ST were particularly poor. While we achieved an improvement in all 11 cases of DT, this was only the case in 5 (38%) of 13 ST patients.

Since 1980, in the surgical treatment of torticollis patients (n=46), those now took place after differential diagnosis of the patient material (ST: n=37, DT: n=9), we achieved the following results:
In ST, we saw 31 (84%) improvements after BMLA, the mortality was 3% (1). As side effects of the procedure we saw in 4 cases a unilateral paresis of the accessory nerve of varying severity. With the stereotactic treatment of DT, 8 (89%) improvements were achieved with tolerable side effects.

Overall, the symptoms improved in 39 of 46 (85%) torticollis patients. The mortality rate was 2%. This corresponds to an improvement in treatment results since the introduction of our new treatment strategy by 19% (see p. 86).
These figures exceed both, the previously published results

with cervical rhizotomy according to FOERSTER/DANDY (up to 75% improvement, 4% mortality, massive side effects: "flappy neck", loss of sensitivity) as well as those achieved with stereotactic brain surgery (improvement 50-60%, mortality 5%, high rate of side effects: hemiparesis, speech disorders, additional peripheral interventions).

Looking at the surgical results of our patients with purely horizontal and purely rotatory symptoms isolated (n=27, 95% improved), these even surpass those of the good results reported by BERTRAND (1987) after selective peripheral denervation (n=111, 87% improved, few side effects).

What striking is the clear difference in the surgical results of patients with purely horizontal and purely rotatoric, i.e. largely limited to the muscles supplied by the acccessory nerve, and patients with combined torticollis and symptoms, which indicate involvement of the deep neck muscles supplied by the cervical plexus. In our experience, a differentiation must be made here, for which speaks the postoperative progression of dystonic symptoms in 4 of 9 patients with mixed torticollis symptoms. According to self-assessment, the symptoms improved in only one of 9 patients. In these patients the presence of a central motor movement disorder now can be assumed and stereotactic surgery must be considered. One of these patients has since undergone successful stereotactic surgery (case 13) (FRECKMANN et al., 1986, 1987).

In contrast to the treatment results with peripheral selective denervation involved in ST symptoms, where we have so far relied exclusively the communication of BERTRAND (1987) himself, the more recent literature already contains reports on the experiences gained by other authors with the BMLA, as publicated by us for the treatment of ST:
In 1983, MOTOMOCHI et al. published a paper on the surgical treatment of ST in conjunction with long-term results. Among other things, they reported on a patient operated using our method with excellent results 7 months after the procedure without any deficit.

Apparently unaware of our work, PAGNI et al. (1985) also published a successfully operated case in which the vascular decompression of the left SANRs had been performed.

In this context, JANNETTA (1986) in the "Journal of Neurosurgery" pointed out our first authorship, through which in the "Neurosurgical Forum - Letters to the editor" of the same journal was triggered a lively response: DYCK (1986) reported a case in which the vascular decompression of both SANRs, firmly anchored to the vertebral arteries, led to a significant improvement of the ST symptoms.

MOTOMOCHI (1986) wrote that JANNETTA had already told him verbally in 1982 of reported 5 good results in a series of 11 ST cases in which the vascular decompression of the SANRs had also been performed. He himself was of the opinion that a considerable number of ST patients could benefit from this operation.

Further 7 cases of ST caused by vascular compression of the SANR were reported by SHIMA et al. (1986, 1987), in whom they had performed BMLA as indicated by us. After a mean postoperative follow-up of 3 years, they saw a complete regression of the ST symptoms in 5 patients and an improvement of symptoms in 2 patients.

In conclusion, it can be stated that the BMLA, after the available long-term observations on our own patient population and with knowledge of the surgical results achieved by other authors, especially in patients with purely horizontal or rotatory ST symptoms that cannot be influenced conservatively, is justified and can be recommended.

If there is no satisfactory improvement in the ST symptoms after BMLA, the treatment proposed by BERTRAND et al. (1981, 1982, 1987), the selective peripheral denervation of individual motor branches of the cervical plexus can be considered. One patient in our group who did not experience sufficient improvement underwent this this procedure 1 year later with good results. The repeated exposure of the SANR,

on the other hand, is only be indicated with great caution due to the increased risk of surgery.

Conclusions

Both our clinical and anatomical findings and our surgical results support the view that ST is not a central motor movement disorder. The cause of ST must therefore be localized peripherally in the craniocervical region.

We assume that the cause of ST is predominantly located in the afferent limb of the head motor system. This is supported by the fact that in our ST patients a frequency of 82% , compared with only 46% of normal sectional specimens where found anastomoses between the SANR and the posterior root C1. This finding indicates an anatomical predisposition that is raised from the latens in middle age and leads to ST symptoms.

In ouer ST patients intraoperatively detected compressions of the SANR and/or the posterior root C1 by the vertebral artery, the PICA or the dentate ligament lead to unilateral irritation of proprioceptive afferents from the head joints and the tension receptors of the musculature supplied by the accessory nerve and to the generation of afferent ectopic impulses, which are processed in a complex manner in the reticular formation. Physiologically, this results in an involuntary contraction of various agonistic neck muscles, which causes a torticollis symptomatology. A secondary central maintenance of the symptoms can be assumed due to the slow postoperative regression.

A peripheral surgical procedure therefore is required for ST. The bilateral elimination of the proprioceptive afferents from the craniocervical region should therefore cause an equalization of the afferent stimulus flow to the formatio reticularis and thus a suppression of one-sided overmotor impulses and a reduction of the cervical muscle tone.
This is the basis of BMLA. Compared to other surgical procedures, BMLA is less destructive, has hardly any side

effects and according to the patients' self-assessment, leads to satifactory results in 95% of the cases with purely horizontal or rotatoric ST symptoms. The different surgical results in patients with horizontal or rotatory ST symptoms and patients with combined torticollis symptoms in the latter indicate the presence of DT with a central motor with central motor genesis.

The indication for BMLA therefore must be limited to horizontal and rotatory ST. Patients with combined torticollis symptoms should be observed as long as possible. In these patients, after confirming the diagnosis of "DT", a stereotactic intervention is recommended.

6.0 Summary

This paper deals the etiologically unexplained picture of the "Spasmodic Torticollis" (ST) with spezial consideration of the particular pathogenesis and surgical treatment. ST is a movement disorder limited exclusively to the cervical musculature, the most obvious symptom is the "torticollis". A causal treatment was not known.

The paper is divided into four parts. In the first part the most important hypotheses regarding the pathogenesis of ST are presented.

In detail, these are
1. the hypothesis of a psychogenic genesis of ST and
2. the hypotheses that assume an organic cause of the desease:
   a) the vestibular/labyrinthine hypothesis
   b) the basal ganglia hypothesis and
   c) the formatio reticularis hypothesis.

These hypotheses have yet to be proven. Apart from exceptions, a psychological cause of ST is not considered probable. In general, the view of a the basal ganglia as the cause of the disease has prevailed. But all neuropathological

findings to date and the results of the surgical treatment speak against a central motor genesis of ST.

In the second part of the study the conservative and surgical methods for the treatment of ST are described and the corresponding treatment results are presented and evaluated.

Regardless of whether in ST only the motoric end-stretch for the neck muscles, such as with cervical rhizotomy, is interrupted or whether stereotactic interventions are performed on the brain, the results are equally unsatisfactory, with a success rate of only 50 to 70%.

Since a causative lesion in the CNS, in contrast to DT, cannot be detected in ST, the lesion must be located peripherally in the area of the motor terminal pathway and/or the proprioceptive afferents responsible for head motor function. These are therefore two etiologically different clinical pictures. Of great importance for the choice of surgical procedure therefore is the differential diagnostic separation of "ST" from "DT".

In the third part of the paper, on the basis of our own anatomical, clinical and electromyographic findings, and of data from 46 torticollis patients who underwent surgery, the theory of a peripheral disorder for the pathogenesis of ST was developed.

In ST patients, 82% of anastomoses are found between the SANR and the posterior root of the first cervical nerve. This is the case in only 46% of the section good. In addition, in 2/3 of the cases a compression of the SANRs with indications of degenerative changes in the anastomotic area, which were confirmed histologically, were found intraoperatively.

The ST therefore is caused by a disturbance of proprioceptive afferents from the head joints and the tension receptors of the muscles supplied by the accessory nerve.
A central maintenance of the symptoms is being discussed.
An anatomical predisposition is assumed, which for

unexplained cause in middle age is brought out of latency.

Our unsatisfactory results with the stereotactic treatment of ST on the basis of this theory made it necessary to develope of a new, peripheral surgical approach.

The BMLA is aimed at the functional elimination of unilaterally disturbed craniocervical proprioceptive afferents in order to achieve a symmetrization of the afferent impulse current from the craniocervical region to the reticular formation and thus cause a one-sided suppression of excessive motor impulses, with a simultaneous reduction of the cervical muscle tone.

After an average postoperative follow-up period of 4 years, the results achieved with the BMLA in 37 patients can be assessed as follows:
In 7 patients there was a complete regression of the ST symptoms, 11 patients had only minor residual symptoms and in 13 patients the result was assessed as improved. In 3 patients, the symptoms remained unchanged and in two patients we noted a deterioration. One patient, after slight improvement, died as a result of a second operation.

When looking at the patients with purely horizontal or rotatory ST symptoms (n=28), it is striking that, with one exception (96%), they had satisfactory results. The poor surgical results in patients with combined torticollis symptoms indicate the presence of an extrapyramidal motor movement disorder. In 4 of these 9 patients postoperatively the symptoms extended beyond the neck.

Compared to other surgical procedures for the treatment of ST, the BMLA is little destructive and has hardly any side effects. Stereotactic brain surgery therefore only should be recommended to patients with proven central motor movement disorders.
This differentiated treatment strategy for torticollis patients leads, as can be shown, to better results than those reported by all previously published statistics.

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PART IV

8.0 Casuistry

Case 1: 42 years old, male, surgery March 1980

Diagnosis: Pronounced horizontal, tonic ST with head turning to the left

Medical history: 1 1/2 years before the operation there was a progressive a compulsive turning of the head to the left with development of severe hypertrophy of the right SCM muscle. Initially, the patient assumed that he had twisted his neck. Every time he tried to turn his head to the right, severe neck pain occurred. All conservative treatment attempts had no effect on the symptoms.

Drug treatment: Akineton, Limbatril, Madopar, Decentan, Tiapridex.

Diagnostics:

Neurological findings: In addition to the pronounced head tilt, there were indications of vestibular irritation on the left with gross nystagmus when looking to the left. ASR attenuated on the right, atrophy and paresis of the right-sided calf muscles (condition after early childhood poliomyelitis).

EMG: In the left trapezius muscle, in the deep neck muscles left and in the right SCM muscle there were strong innervations, which could still be broken arbitrarily. No signs of tremor.

CT: No evidence of pathological changes.

Vertebral angiogram: unremarkable findings.

EEG: Alpha type without focal findings, no cramp potentials.

X-ray: Malposition of the cervical spine with degenerative changes and slight narrowing of the intervertebral foramina between cervical vertebrae (CV) 5 and 6. No evidence of a vertebral cause of the torticollis.

Operation: After exclusion of a torsion dystonia or another extrapyramidal disease, right dorsolateral exposure of the posterior cranial fossa up to the spinal process CV2. Osteoclastic trepanation of the right posterior fossa. Opening of the occipital foramen magnum. Resection of the dorsolateral right atlas arch. Opening of the dura and exposure of the right SANR.

The accessory nerve is connected to the dentate ligament and the vertebral artery at its passage through the dura. The posterior spinal artery emerges from the adhesion cord. The adhesions are sharply separated, the right accessory nerve is completely detached from its adhesions and preserved. The nerve itself appears inconspicuous. Padding of the accessory nerve with a Prosthex sponge. A dorsal C1 root is not present corresponding to type 1. Wound closure.

Postoperative course: After the operation there was extensive regression of the ST symptoms and normalization of the tone in the right SCM and trapecius muscles. 2 weeks after the operation sudden deterioration with fixed ST to the left. 6 months after the operation gradual regression of the symptoms. EMG revealed improved findings on the right side with moderate reparative processes. On the left signs of old neurogenic damage to the SCM muscle. Further follow-up examinations finally revealed complete freedom from symptoms and complaints.

Case 2: 21 years old, male, surgery April 1980

Diagnosis: Pronounced horizontal ST to the left

Medical history: 1 1/2 years prior to the surgical treatment an involuntary tilting of the head with a turn to the left. In the further course, increase in torticollis symptoms with complete fixation of the head in the incorrect posture and hypertrophy of the right SCM muscle. Finally, the head was turned 90 degrees to the left and fixed in this position. At maximum effort, the head could only just be brought to the center position.

Drug therapy: Diazepam, carbamacepine, Tiapridex, Dartal.

Diagnostics:

Neurological findings: Apart from the ST symptoms, the patient's status was downright.

EMG: In the left SCM and trapezial muscles increased discharge frequencies and relatively frequent neurogenic action potentials, which were indicative of damage to the left accessory nerve. The assessment of individual action potentials was difficult due to bursty innervations. Minor involvement of the deep neck muscles. Inconspicuous findings on the right side.

CT: Inconspicuous intracranial findings.

Vertebral angiogram: Atypical course of the left PICA, which crosses the midline with a loop about 4 mm to the right.

EEG: Basic activity in the alpha range, the course of the curve appears somewhat more irregular in the parasagittal derivation on the right than on the left.

Surgery: After exclusion of torsion dystonia or another extrapyramidal disease, depiction of the right SANR. It is fused with the ligamentum denticulatum below CV1. At the level of dura passes through of the vertebral artery, the SANR is taut over the vertebral artery and the posterior spinal artery that branches off there at the same time. There is an anastomosis between the SANR and the posterior root C1,

corresponding to type I. In addition, the posterior root C2 is connected to the caudal branch of the posterior spinal artery. After severing of the uppermost denticular ligament at the level of the foramen magnum, the accessory nerve is separated from its ligament, as well as the vertebral artery and the posterior spinal artery. The posterior root C1 is severed at its dural entry. Due to the tight devouring of the posterior root C2 with the spinal artery, this is also severed. Finally, a prosthex implant is placed between the accessory nerve and vertebral artery and the wound closed.

Postoperative course: Initially only slight improvement in the symptoms of torticollis. However, the head arbitrarily could now be brought into a central position. In the course of the rehabilitation treatment, further improvement was achieved through biofeedback training and ST gymnastics. 9 months postoperatively, a further regression of the symptoms occurred. Electromyography showed an extensive regression of the changes observed before the operation. 7 years after the operation there existed only residual symptoms with largely restored voluntary motor function.

Case 3: 47 years old, male, surgery in July 1980

Diagnosis: Horizontal ST with head turned to the right

Medical history: One year before the operation development of a ST, that could not be influenced by medication, with largely fixed head turned 80 degrees to the right. Arbitrary turns to the left were only possible up to 45 degrees. Significant easing facilitation of voluntary head motor skills with the help of the “Geste antagoniste”.

Drug therapy: Tiapridex, Decentan, Akineton, Saroten.

Diagnostics:

Neurological findings: Apart from the ST symptoms and a high degree of hypertrophy of the left SCM muscle, unremarkable

findings without indication of torsion dystonia or another extrapyramidal disorder.

EMG: The splenius capitis, trapecius and SCM muscles were active on the right side. There were evidence of older neurogenic damage in the left SCM muscle, to a lesser extent also in the left trapezial muscle.

CT: Intracranial no pathologic findings.

Vertebral angiogram: The left vertebral artery forms a cranial loop just above the level of the foramen magnum. Extremely deep exit of a large caliber PICA, which forms a loop reaching far caudally below the foramen magnum and extending far to the right.

X-ray: Slight narrowing of the intervertebral disc between CV6/7 with slight spondylotic edge protrusions without constriction of the intervertebral foramina.

EEG: Inconspicuous alpha EEG.

Operation: BMLA performed for the first time as described.

Surgical findings: On the right side, the SANR is stretched over the PICA, which also descends deep and is adherent to it. The dorsal C1 root corresponds to type III. An anastomosis exists between the accessory root and the posterior C1 root, which has a ganglion. In addition, there is a distortion of the accessory nerve root with torsion by 180 degrees, about 5 mm above the adhesion site. At the slightest touch to the accessory nerve leads to contraction of the right SCM muscle.

On the left there is a very deep exit of the PICA from the vertebral artery even before it passes through the dura. The PICA forms a caudally directed and in a large arc dorsal to the dorsal of the upper cervical medulla up to above the midline and then bends in a cranial direction. The left SANR is clearly stretched over the PICA. The left posterior C1

root corresponds to type IV. It connects anastomotically with the accessory nerve.
An EMG recording was made intraoperatively after electrical stimulation of individual accessory nerve root threads and the cervical posterior roots.

Both accessory nerve roots are removed from all adhesions with the denticular ligament, the dorsal C1 roots and the PICA are detached. The anastomosis between the posterior root C1 and the accessory nerve is preserved on both sides. Then accessory nerve roots are protected from renewed vascular contact by Prosthex implants.

Postoperative course: Initially only slight improvement. Under intensive physiotherapy, the symptoms improved to such an extent that the patient could voluntarily hold his head straight. After gradual regression of symptoms, there was a further deterioration 6 months postoperatively, finally to a complete recurrence. Compared to the preoperative symptoms, however, the patient no longer had any pain.

On the assumption that either the implants could have dislocated, or that scarred adhesions with renewed irritation of the SANR, it was decided to revise the surgical site. This was performed 15 months after the first operation. After removal of extensive scarring and adhesions, the posterior roots C1 and C2 now was severed.

Postoperatively, there was an immediate improvement in the voluntary motor function of the head. The symptoms of torticollis regressed. As a result of the procedure, there was a circumscribed analgesia emphasized on the right in the area supplied by C2 and a slight bilateral accessory nerve palsy. In the further course the condition stabilized and improved significantly. 6 years after the 2nd operation,there was still a slight turn of the head by 20 degrees to the right. The accessory nerve palsy had regressed. The patient was already retired since the first operation and was satisfied with the condition achieved.

Case 4: 46 years old, female, surgery in October 1980

Diagnosis: Position-dependent horizontal ST to the right

Medical history: 2 years before the operation the patient for the first time noticed an involuntary turn of the head to the right. The symptoms were particularly pronounced when lying down and leaning the head. This was accompanied by clonic turning movements of the head to the right, in conjunction with severe neck pain.

Drug therapy: Akineton, Tiapridex and Dartal.

EMG: With involvement of the SCM and trapecius muscels on both sides, more pronounced on the left than on the right, indications for partially old or very old but also for for more recent neurogenic lesions were found.

CT: No pathologic changes intracranially.

Vertebral angiogram: unremarkable findings.

Surgical findings: BMLA; there is an anastomosis between the SANR and the posterior root on the right side C1 corresponding to type IV. A root fiber of the posterior root C2 has a common course over a short distance with the SANR and enters the spinal cord below the level of C1. There are close neurovascular contacts between the accessory root and the vertebral artery.

There are also anastomoses on the left between the SANR and the posterior roots C1 and C2. The anastomosis with C1 corresponds to type III. The SANR is stretched over the denticulate ligament. The posterior spinal artery that departs from the dural passage of the vertebral artery and its branches is wound several times around the SANR.

Postoperative course: The ST symptoms were already significantly improved on discharge. After rehabilitative treatment, the ST symptoms regressed further with

stabilization of the condition. 6 years after the operation, a good result with discrete residual symptoms was observed.

Case 5: 52 years old, male, surgery in October 1980

Diagnosis: Horizontal ST to the left with “Geste antagoniste”

Medical history: 4 years prior to the operation, the patient developed progressive horizontal ST with head turning to the left. The disease initially began with a feeling of tension in the left neck region. A short time later the patient noticed that the head was turning more and more involuntarily to the left. However, it was possible to return the head to a normal position. In the further course of time symptoms intensified and there were sometimes very violent, clonic rotations of the head. With help of the “Geste antagoniste", the patient was able to influence the symptoms favorably. When the right hand was removed, the head immediately turned 90 degrees to the left.

Drug therapy: Haldol, Akineton, Cosaldon, Tavor, Cortisone and Dartal.

Diagnostics:

Neurological findings: Apart from the ST symptoms with hypertrophism of the right SCM muscle, the neurological findings were were normal. There were no indications for torsion dystonia or any other extrapyramidal disorder.

EMG: In the SCM and trapezius muscles more pronounced on the right, clear evidence of neurogenic damage. No signs of damage in the splenius capitis muscle on both sides.

CT: Intracranial no pathologic findings.

Vertebral angiogram: Unremarkable findings.

EEG: Pathological changes are not detectable.

Operation: BMLA; On the right there is an anastomosis between the SANR and posterior root C1, corresponding to type IV. The anastomosis is stretched over the upper edge of the denticulate ligament. At the level of C2, the accessory nerve runs below the sensitive C2 root also taut over the C2 root artery. In the anastomosis area a ganglion can be seen.

On the left, the SANR is connected to a strong loop of the posterior spinal artery. A dorsal C1 root is not found, corresponding to type I. However, there is an anastomosis between the accessory nerve root and the dorsal C2 root.

Postoperative course: After the operation the torticollis symptoms improved only slightly. A significant improvement, which was also satisfactory for the patient did not occurred. The patient died 4 years after the operation in status asthmaticus with pre-existing bronchial asthma.

Case 6: 22 years old, male, surgery February 1981

Diagnosis: Horizontal ST with head turned to the right

Medical history: 5 years before the operation the patient suffered an accident with brain contusion and unconsciousness for three weeks. One year later sudden onset of ST symptoms. There was an increasing tilting of the head with turn to the right. Since then this remained constant. All conservative treatment attempts were unsuccessful.

Diagnostics:

Neurological findings: In addition to the ST with hypertrophy of the left SCM muscle, there was an incomplete hemianopsia to the right as a result of the brain contusion. Signs for the presence of torsion dystonia or another extrapyramidal disease were not found.

EMG: Clear evidence of neurogenic damage in the left SCM muscle.

X-ray: Significant scoliosis with left deflection of the cervical spine.

CT: Intracranial no pathological findings. Signs of an older fracture of the posterior arch of the atlas.

Vertebral angiogram: Deep exit of the right caliberstrong PICA.

Operation: On the right there are close neurovascular vascular relationships between the SANR and the PICA. Through a dorsally directed loop of the PICA, the accessory nerve is clearly tensed. In addition the SANR is adhaerent with the caliberstrong vessel. There are also 2 fine anastomoses of the the accessory root to the delicate posterior root C1 and to the posterior root C2, corresponding to type III.

On the left there are inconspicuous relationships, the dorsal C1 root corresponds to type II.

The histological examination of the posterior root C1 and the posterior root C2 revealed no special features.

Postoperative course: Immediately after the operation the patient experienced an improvement in his symptoms, although objectively no change in the symptoms could be recognized. 6 months after the operation the symptoms gradually regressed to a large extent. A slight posture of the head, which only slightly impaired the patient's ability to work remaind.

Case 7: 36 years old, male, surgery February 1981

Diagnosis: Horizontal ST to the left

Medical history: 4 years before the operation, a rapidly

developing, increasing forced posture of the head with a turn and slight inclination to the left. At the same time a marked hypertrophy of the left SCM muscle developed.

Drug therapy: Tavor, Decentan, Lioresal, Artane, Tiapridex, Limbatril, Muscle Trancopal and Dartal.

Diagnostics:

Neurological findings: Massive turning and slight inclination of the head to the left. Left shoulder elevation. Significant increase in symptoms under emotional influences. Aside from that, a bending posture and abduction of the left arm was noticed when walking, so that a dystonic torticollis was considered. Otherwise no neurological abnormalities. On the basis of the angiographic and electromyographic findings, the decision nevertheless was made to perform BMLA, although in this case the presence of torsion dystonia could not be completely ruled out.

EMG: Emphasized in the right SCM muscle there are partially reduced, partially enlarged, frequent discharging action potentials. The findings suggest chronic neurogenic damage to the right accessory nerve.

CT: unremarkable findings.

Vertebral angiogram: PICA outgoing relatively high in a loop, with otherwise unremarkable findings.

EEG: Laterally symmetrical alpha sequence without special features.

Operative findings: On the right side there was an anastomosis between the SANR and the posterior root C1, corresponding to type IV (Fig. 11).

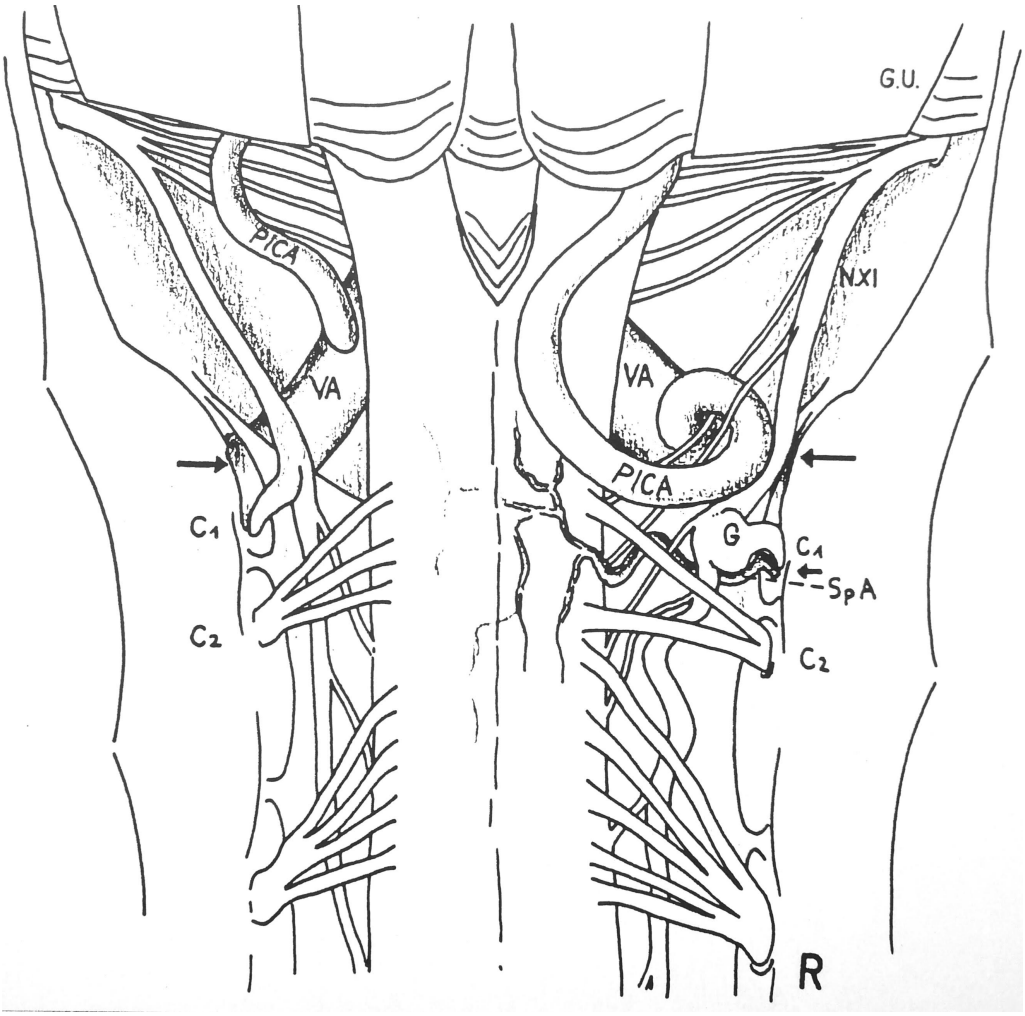


Fig. 11: Case 7, Operations sketch

On the left, there is also an anastomosis between the accessory nerve and posterior root C1, corresponding type IV. In the anastomosis area a small ganglion can be recognized on the dorsal C1 root, transitioning the SANR. It is firmly fixed to the dorsal C1 root and to the dentate ligament. In addition, the SANR is pressed trough the PICA sling against the edge of the foramen magnum. The intraoperative electrical stimulation with electromyographic recording indicates efferents in the posterior root C1. There is a contraction of the left SCM muscle.

Postoperative course: An improvement in symptoms could be seen immediately after the operation in that the patient can

turn his head more easily to the right. In the further course, after about 6 months there was initially a good regression of the torticollis symptoms. 1 1/2 years after the operation, however, there was a slight deterioration so that today only a slight improvement of the ST symptoms can be noticed.

Case 8: 26 years old, female, surgery May 1981

Diagnosis: Pronounced horizontal ST to the left with “Geste antagoniste”

Medical history: 21 months before the operation, the patient had suffered a hit on the right shoulder with an iron bar. One week later, an increasing ST with turning of the head to the left. In the further course, the patient developed an “Geste antagoniste”, touching with his left hand on the left chin. At the same time, the right SCM muscle hypertrophied. A clear positional dependence of the torticollis was indicated. In the supine position the head could be moved freely, the forced posture was lifted. Conservative therapy led to a further intensification of the symptoms.

Drug therapy: Dartal, Akineton.

Diagnostics:

Neurological findings: Apart from the predominantly tonic ST symptoms, normal findings.

EMG: Fresh neurogenic damage in the right SCM muscle.

CT: No pathological findings intracranially.

EEG: Inconspicuous brain waveform.

Vertebral angiogram: Deep exit of the PICA from the right vertebral artery below the level of the foramen magnum.

Surgical findings: On the right side there is an anastomosis between the posterior root C1 and the SANR corresponding to type IV. The SANR is displaced, tensed and compressed medially by a strongly tortuous PICA that runs deep along the dural passage of the vertebral artery. The course of the SANR is forced by a connected very short posterior root C1. In the area of the anastomosis a ganglion is found.

On the left there is also a type IV anastomosis, with a spinal ganglion C1 located in the anastomosis area. Here the SANR is slightly raised by a small spinal artery, that emerges from the vertebral artery.

In typical manner, both accessory roots were released from their vascular contacts. The anastomoses to the posterior roots C1 are severed. The accessory roots are underpadded with Prosthex.

Postoperative course: A clear regression of the head misalignment was observed relatively quickly. Initially the patient stated that he had no feeling for the head position. With exercises in front of the mirror he was able to correct the head position. At the time of the follow-up examination 67 months after the operation, the ST symptoms were completely regressed and the patient was symptom-free. The symptoms regressed 18 months after the operation.

Case 9: 42 years old, male, surgery September 1981

Diagnosis: Horizontal ST with clonic turning movement of the head to the left

Medical history: 10 years before BMLA increasing development of a ST with unsuppressible clonic turning movements of the head to the left. 2 1/2 years later intensification of the symptoms with hypertrophy of the right SCM muscle. Conservative treatment measures including psychotherapy did not lead to any improvement. After a further, considerable increase in symptoms, 6 years after the onset of the disease,

a stereotactic thalamo- and subthalamotomy was performed on the left side. A change in the symptoms could not be achieved. Further conservative treatment attempts did not lead to any improvement either. Over the course of the years, the patient developed the “Geste antagonist”, with touching the left cheek with the left hand.

Drug therapy: Tiapridex, Decentan, Lioresal, Artane, Dantrium.

Diagnostics:

Neurological findings: In addition to the ST symptoms, there were discrete right leg hemiparesis as a residual finding after thalamo- and subthalamotomy.

EMG: Clear evidence of neurogenic damage to the right-sided muscels supplied by the accessory nerve.

CT: Inconspicuous intracranial findings.

Vertebral angiogram: No deviations from the norm.

EEG: Unremarkable curve progression.

Surgical findings: Type IV anastomosis on the right side. The SANR is pulled into the vertebral foramen C1, there adhaerent and clearly tense. In addition the SANR is in close contact to a spinal artery that arises directly from the dural passage of the vertebral artery.

On the left there is also a type IV anastomosis. The SANR is connected to the spinal vertebral artery at their dural passage. In addition, there is a ganglion on the dorsal root C1 in the anastomotic area.

Histologic findings: In the left posterior root C1 there is a small but clearly recognizable Schwann-cell proliferation. In addition, some axons are drifted. This finding indicates

minor de- and regeneration processes.

Postoperative course: As a result of the operation a moderate paresis of the lateral elevation of the right arm developed. 5 1/2 years after the operation, the ST symptoms have largely regressed, the patient has full control over the voluntary motor function of the head.

Case 10: 25 years old, female, surgery February 1982

Diagnosis: Horizontal ST with head-turning to the right

Medical history: 1 1/2 years before the operation, during a carousel ride a “dislocation” of the cervical spine occurred. The emergency doctor had performed a cervical spine adjustement. A few days later, the head began to turn involuntarily to the right. Temporarily, the medication and physiotherapy treatment led to an improvement. However, the symptoms ultimately remained unaffected and increased in severity. The symptoms temporarily could be favorably influenced by the “Geste antagoniste”.

Drug therapy: Tiapridex, Dartal.

Diagnostics:

Neurological findings: Apart from the ST symptoms regular status.

EMG: The findings suggest neurogenic damage to the muscles supplied by the accessory nerve, more pronounced on the on the right than on the left.

CT: No pathological findings intracranially.

Vertebral angiogram: Inconspicuous vascular image.

EEG: Inconspicuous brain waveform.

VEP: No definite pathological findings.

Vestibular examination: Calorically equally excitable on both sides, position-dependent nystagmus when turning the head to the right.

Surgical findings: Type IV anastomosis on the right side without vascular nerve contacts.

On the left side there also is a type IV anastomosis with the posterior root C1, which contains a rice-sized reddish injected ganglion to the SANR. The ganglion is dissected from the SANR and resected. Also there is a close contact between the SANR and the vertebral artery, which is released.

Postoperative course: Initially no significant change in symptoms. 12 months after the operation there was a significant improvement of the symptoms, at times there was complete freedom from symptoms. Later we were unable to follow up with the patient as she had moved away.

Case 11: 41 years old, male, surgery March 1982

Diagnosis: Pronounced horizontal ST to the left

Medical history: 14 months before the operation the patient developed a pronounced ST with head turning to the left. When lying down improvement of the symptoms. When standing and walking the head was turned 90 degrees to the left and could be move to the center position only up to about 30 degrees. At the same time, an unusually strong hypertrophy of the right SCM muscle and the left trapezial muscle developed.

Therapy: biofeedback therapy, local procaine injections, Tiapridex, Dartal, physiotherapy.

Diagnostics:

Neurological findings: Apart from the pronounced ST-symptoms, unremarkable findings.

EMG: Clear evidence of neurogenic damage to the right SCM muscle.

CT: Unremarkable findings.

Vertebral angiogram: Low level and pronounced slenderness of the right PICA.

Surgical findings: On the right side there is an anastomosis corresponding type IV. The SANR is inserted into the intervertebral foramen C1 and there fixed on the vertebral artery. Besides there is a ganglion in the anastomosis area.

On the left anastomosis type III. The posterior root C1 has a common course here over 15 mm with the SANR.

Postoperative course: Initially,despite easing of the tension SCM muscle, no significant change in ST symptoms. 5 years after the operation very good regression of the ST symptoms.

Case 12: 50 years old, male, surgery March 1982

Diagnosis: Horizontal ST with head turned to the left

Medical history: 13 years before the operation, ST symptoms developed increasingly with involuntary turning of the head to the left. Two years later, the patient suddenly developed a permanent fixed head misalignment. With Haldol, an improvement was initially achieved for 6 months. In the following years the patient was treated with Dartal and Akineton. A remission was achieved which lasted for about 5 years. Two years before the operation, a relapse occurred. The neurological findings at this time revealed an anisocoria with a wider pupil on the right side and a right-sided increase of the muscle stretch reflexes. The abdominal skin

reflexes were weaker on the right than on the left. The repeated medicamentary treatment led to considerable side effects with further development of the ST symptoms.

Drug therapy: Akineton, Dartal.

Diagnostics:

Neurological findings: Apart from the ST symptoms, the neurological findings were completely normal before the operation. The right SCM muscle was clearly hypertrophied. No reference to a torsion dystonia.

EMG: Indication of neurogenic damage to the muscles supplied by the accessory nerves, more pronounced on the right than on the left.

CT: Inconspicuous intracranial findings.

Vertebral angiogram: Loop formation of the left PICA, extending far caudally, otherwise unremarkable findings.

EEG: Inconspicuous brain waveform.

Operative findings: On the right side SANR anastomosis correspondig type IV without direct connection between the posterior root C1 and the spinal cord. Some root fibers of the posterior root C2 move cranially into the area of the normal C1 entry zone. There is a neurovascular contact to a dorsal spinal artery, that originates at the dura-passage of the vertebral artery and crosses under the accessory nerve.

On the left there is a SANR-C1 anastomosis type III. There is also a close neurovascular contact to a caliber strong dorsal spinal artery originating at the dural passage of the vertebral artery. A further anastomosis exists between the posterior root C2 and the SANR.

Histologic examination: Normal structure of dorsal root parts with nerve fiber bundles and ganglion cells.

Postoperative course: Postoperatively, there was a good regression of the ST within 3 months. Almost 5 years after the operation, the patient's condition can be described as very good, he is fully employed and practically symptom-free.

Case 13: 47 years old, male, surgery April 1982

Diagnosis: Pronounced horizontal ST with head turning to the right.

Medical history: 1 1/2 years prior to the surgical treatment without any recognizable external cause progressive development of a ST symptomatology. Practicing the medical profession became impossible. Although there was initially a slight improvement in the symptoms under conservative treatment, the patient insisted on undergoing BMLA.

Conservative therapy: medication with Dartal and Akineton, physical therapy with physiotherapy and exercise baths.

Diagnostics

Neurological findings: Although not definitely nystagmus, jerky movements of the eyeballs were noticeable. Otherwise, downright apart from the ST symptoms and a questionable increase in the tone of all extremities downright.

EMG: Minor neurogenic damage on both sides, predominant on the right side damage in the SCM and trapezius muscles.

CT: Inconspicuous intracranial findings.

Vertebral angiogram: normal findings.

Surgery: Type IV anastomosis on the right side. The C1 ganglion is firmly attached to the SANR and the entire complex is retracted into the foramen C1 and adherent here. In addition, there is an anastomosis between the SANR and the posterior root C2.

On the left there also is a type IV anastomosis. Here a vascular-nerve contact exists between the PICA and the SANR, which is compressed by a PICA loop and is pressed onto the denticulate ligament.

Postoperative course: Immediately after the operation that the involuntary right rotation of the head had disappeared. In the further course the ST symptoms improved significantly within 6 months. The patient complained that his head was still a little wobbly, but that he could control arbitrarily. He had meanwhile resumed work. Nevertheless, the patient was still not satisfied with the result of the operation, so that he contacted Prof. Bertrand in Montreal 1 year after the operation. There, the peripheral selective denervation was performed on both sides of the neck. The procedure initially was to the patient's complete satisfaction. 3 years later, however, he had a second operation in Canada. According to telephone information, he is doing very well again.

In this case, it should be noted that the patient with mild ST symptoms was very quickly to undergo surgery, even though he was aware of the limited chances of successful surgery to treat ST.

Case 14: 27 years old, male, surgery June 1982

Diagnosis: Horizontal ST with slight head tilt to the right

Medical history: 2 1/2 years before BMLA, pain initially occurred in the cervical spine area. 3 months later ST symptoms developed progressively with head turning to the right. 2 years before surgery, in addition to a head tremor which accompanied the head malposition, a reduced movement of the right arm when walking was observed. At the same time involuntary non-rhythmic movements of the right arm. The first EMG analysis from the left SCM muscle revealed a regularly tremor with a frequency of 10/sec. The medication

treatment with Tiapridex and Limbatril resulted in a temporary improvement of the symptoms, but the cervical spine pain persisted. In the further course of the disease, which no longer responded to conservative treatment measures, the head malposition was largely fixed.

Drug therapy: Tiapridex, Limbatril, Akineton, Dartal and Muskeltrancopal

Diagnostics:

Neurological findings: The head was in a constant malposition with a pronounced turn to the right. Active and passively, the head could be brought to a maximum in the middle position. The remaining neurological findings now were normal compared to the initial examination. Indications of an extrapyramidal movement disorder were not recognizable.

EMG: Minor neurogenic damage in the SCM muscle, less also in the splenius capitis and trapecius muscles on the left. Unremarkable findings on the right side.

EEG: normal findings.

CT: Normal brain wave curve.

Vertebral angiogram: normal visualization of the vertebral artery area.

Surgical findings: There is a type IV anastomosis on the right and additionally an anastomosis of individual root fibers of the posterior root C2 to the SANR. One root fiber of the posterior root C2 extends cranial into the entry area that is normally assigned to the root C1. The SANR has a vascular-nerve contact with a posterior spinal artery that originates at the passage of the vertebral artery.

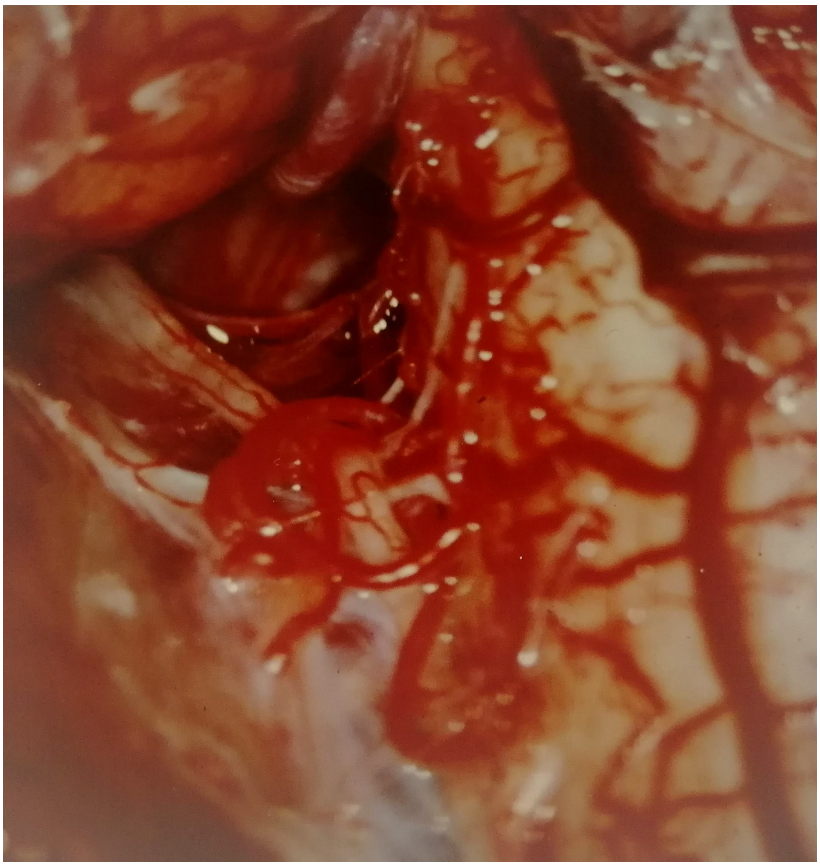


Fig. 12: Case 14, left side

On the left there is a type III anastomosis. A root fiber C1 reaches its normal entry area at the brain stem. There is a close neurovascular contact of SANR with a posterior spinal artery, which also originates directly at the dural passage from the vertebral artery. This artery forms a loop that is firmly attached to the accessory nerve root (Fig. 12).

Postoperative course: Postoperatively there was hardly any improvement initially observed. In contrast, slight perioral dyskinesia and a fine tremor at all extremities were observed. Five weeks after the operation developed intracranial pressure symptoms with nausea, vomiting and mental changes. In addition, there was a clear cerebellar ataxia. The control CT scan showed enlarged ventricles, so that a lumbar cerebrospinal fluid pressure measurement was carried out, which elevated borderline CSF pressure values.

A ventriculo-atrial shunt should be placed. After removal of

the lumbar catheter, however, there was a rapid spontaneous improvement in the cerebellar symptoms and a regression of the headache. At the same time, there was both subjectively and objectively an improvement of the ST symptoms. In the further course, however, there was no further significant improvement of the ST symptoms. Even today, compared to the pre-operative state, only a slight regression of the head malposition occurred, so that further attempts with drug treatment with Xylotocan and Akineton were carried out. This, too, had not yet a satisfactory result.

Case 15: 38 years old, female, surgery August 1982

Diagnosis: Horizontal ST with head turned to the right

Medical history: 1 1/2 years before the operation, after initial pulling pain on the right side of the neck region, a horizontal ST with a small retrocollis component developed. Conservative treatment measures did not lead to an improvement. The involuntary turning of the head to the right was more pronounced when sitting than when standing and lying down. However, under great effort the patient was able to turn her head to the left. Gradually further deterioration occurred. The head almost was fixed in the wrong position, accompanied by tremor-like restlessness of movement.

Conservative therapy: acupuncture, medication with Akineton, Tiapridex, Dartal.

Diagnostics:

Neurological findings: Apart from the ST symptoms, the neurological findings were normal, in particular there were no signs of a central movement disorder. The left SCM muscle was clearly hypertrophied.

X-ray: Left deflecting torsion scoliosis of the cervical spine.

CT: Slight enlargement of the lateral cerebral ventricles and atrophy in the area of the insular cortex. Otherwise unremarkable intracraial findings.

EMG: Clear signs of right-sided neurogenic damage in both SCM muscles.

Vertebral angiogram: Deep exit of the right PICA at the dural passage of the vertebral artery with pronounced tortuosity.

Surgical findings: Right side corresponding to type II. The posterior root C2 showed slight adhesions to the SANR without any anastomosis. The posterior root C1 enters the lateral posterior sulcus at a normal position. As recognizable in the angiogram, there was a deep outlet of the PICA from the vertebral artery at the dural passage. The SANR runs in close contact to the vertebral artery through the bifurcation, which is formed from the vertebral artery and PICA.

On the left, the conditions correspond to type III, the posterior root C1 maintains an anastomosis with the SANR. In the area of the anastomosis, the SANR is drawn into the intervertebral foramen C1 and is adherent there. Medially, the posterior root C1 reaches its normal entry field into the medulla. In addition, there is an anastomosis between the posterior root C2 and the accessory root.

Postoperative course: Initially, the patient only noticed a slight reduction in involuntary head movement to the right. 6 months after the operation, there was a gradual almost complete regression of the ST symptoms. The pain also subsided. The patient was able to return to work. In the meantime, the treatment result can be considered very good.

Case 16: 47 years old, female, surgery March 1983

Diagnosis: Horizontal ST with head turned to the right

Medical history: 3 1/2 years before the operation, following a severe febrile tonsillitis the development of a horizontal ST with turning of the head to the right. Initially the symptoms were discrete, the patient was first made aware of the head malposition by a friend. The patient learned to control the symptoms with the help of the “Geste antagoniste”. In the further course a progression of the symptoms with fixation of the head misalignment occurred, so that the head finally could only be brought into the middle position. The ST symptoms increased under emotional stress, while lying down or leaning the head an improvement occurred. In addition, the patient complained of severe neck pain, occasionally radiating into both shoulders. The patient also noticed a feeling of tension in the left hypertrophied SCM muscle. Conservative treatment measures were unable to achieve an improvement.

Conservative therapy: physiotherapy, exercise baths, hypnosis, medication with biofeedback training, Tiapridex, Limbatril, L-tryptophan.

Neurological findings: Except from the ST symptoms with hypertrophism of the left SCM muscle, the neurological findings were normal.

EMG: Neurogenic damage in the muscles supplied by the accessory nerve on both sides, more pronounced on the left.

EEG: Inconspicuous brain waveform.

CT: Moderate enlargement of the 3rd ventricle, enlargement of the insular cisterns, suspected upper worm atrophy.

Vertebral angiogram: Deep exit of the right PICA with formation of a deep loop up to the level of the atlanto-occipital ligament. On the left side there is also a deep descending loop of the PICA.

Surgical findings: The right side corresponds to type III. The dorsal C1 root reaches the spinal cord at a normal

position with an anastomosis to the SANR. The angiographically abnormally deep PICA loop with deep exit from the vertebral artery presses the SANR onto the vertebral artery. The SANR is fixed here by arachnoid adhesions.

On the left, there is also an anastomosis corresponding to type III. Here, too, the dorsal C1 root reaches the vertebral artery after formation of an anastomosis. The deep-reaching PICA loop is in contact with the SANR.

Postoperative course: Initially, recovery was characterized by persistent neck and forehead headaches. Then there was improvement of the torticollis symptoms, which was strongly dependent on the intensity of psychological stress. 6 months after the operation, the improvement in head misalignment was clearly recognizable. The head could now be held in the middle position again and also be turned to the left. The neck pain had disappeared. The treatment result can be described as good.

Case 17: 45 years old, male, surgery May 1983

Diagnosis: Horizontal ST with head turned to the right

Medical history: 2 years before the operation, a slowly progressive the development of ST symptoms with turning of the head to the right. With the help of the “Geste antagoniste” it was initially possible to influence the symptoms favorably. Finally, a massive head misalignment developed, associated with a clonic restlessness of movement of the head. The symptoms were more pronounced when standing and walking than when lying down. All conservative treatment measures were unsuccessful.

Conservative therapy: physiotherapy, biofeedback training, Haloperidol, Tiapridex, Akineton, Rivotril, Dartal.

Neurological findings: In addition to the ST symptoms, there was a hypertrophy of the right SCM muscle. Sensitivity

disorder on the ulnar side of the left hand, as well as on both outer edges of the foot in a condition following surgical removal of a herniated lumbosacral disc. The remaining neurological findings were normal.

EMG: In the right SCM muscle there were evidence of older neurogenic damage, less pronounced also in both trapezius muscles.

EEG: Inconspicuous brain waveform.

CT: Inconspicuous intracranial findings.

Vertebral angiogram: Inconspicuous visualization of the vertebral-basilar vascular system.

Surgical findings: On the right side, the relationship between the SANR and the posterior root C1 corresponds to type III. There are no vascular-nerve contacts.

On the left side, the relationship between the SANR and the posterior root C1 corresponds to type IV. The posterior root C1 enters the spinal canal with the vertebral artery and forms a yellowish-colored ganglion-like structure, which connects to the SANR and ends here. The accessory root is firmly attached to the vertebral artery. In addition, a posterior spinal artery arises from the dural passage of the vertebral artery, which originates in the area of the anastomosis between the SANR and C1 and also forms a vascular-convolute with contact to the accessory nerve root.

Postoperative course: Soon after the operation, the patient noticed a regression of the neck pain. The ST symptoms appeared slightly improved, the patient was able, albeit with effort, to turn his head beyond the center to the left. In the further course the symptoms regressed very well and the patient was able to resume work. The treatment result can be described as very good.

Case 18: 57 years old, male, surgery April 1984

Diagnosis: Horizontal ST with head turned to the left

Medical history: 7 years before the operation, the patient noticed first noticed involuntary, initially only at night occurring turning movements of the head to the right. Soon the symptoms also occurred during the day. Later, there was a change in the direction of rotation of the head, the patient could no longer remember the exact time. A violent involuntary pulling of the head to the left developed. Conservative treatment measures had no influence on the symptoms. An arbitralily turn of the head to the right finally became almost impossible. With the help of the “Geste antagoniste”, the patient temporarily was able to influence the symptoms favorably. At the same time, however tremor-like restlessness of movement of the head apeared.

Drug therapy: Gutron, Tiapridex, Tremarit, Dartal.

Diagnostics:

Neurological findings: Except for the ST symptoms with hypertrophism of the right SCM muscle and a fasciculation in the upper trapezius portion on the left, regular neurological findings. In particular, there were signs of an extrapyramidal motor disorder.

EMG: There were slight indications of neurogenic damage in the muscles supplied by the right accessory nerve, particularly the SCM muscle.

CT: Inconspicuous intracranial findings.

Vertebral angiogram: There is a deep PICA loop, under the level of the foramen magnum on the left side. The origin of the PICA lies at the dural passage of the vertebral artery.

Surgical findings: On the right side, the conditions

correspond to type I. There is no dorsal C1 root. The SANR is completely uninhibited and normal.

According to type I, there is no posterior root C1 on the left side. The PICA loop recognizable in the angiogram has its origin at at the passage of the vertebral artery. It forms a loop directed posteriorly and caudally, then pulls cranially into the cerebellopontine angle. There it forms another loop caudally, which continues below the level of the foramen magnum. The left SANR root is clearly extended over the PICA in the area of its origin from the vertebral artery. After this, the accessory root has a common course with the PICA for about 1.5 cm. The accessory root is firmly fused with the PICA. This is a pronounced neurovascular contact.

Postoperative course: After the operation innervations in the right SCM muscle were no longer observed. 6 months after the operation the ST symptoms had largely regressed, the patient now was able to move his head freely and arbitrarily in all directions. There was only a slight tremor-like restlessness of the head posture. The patient had resumed his work. The treatment result can be described as good.

Case 19: 41 years old, male, surgery in June 1984

Diagnosis: Horizontal ST with head turned to the right

Medical history: 9 months before the operation there was a rapid increase involuntary turning of the head to the right. Occasionally there were also shooting innervations with clonical restlessness of the head and slight retrocollis symptoms were observed. The most pronounced symptoms were when standing and walking, while the symptoms almost disappeared in the supine position. With the help of the “Geste antagoniste”, the patient was able to influence the symptoms favorably. Conservative treatment measures did not lead to any improvement of the symptoms.

Conservative therapy: physiotherapy, exercise baths,

bio-feedback training, medication with Akineton, Tiapridex, Dartal, Xylotocan.

Diagnostics

EMG: Indications of slight chronic damage to accessory nerve on the left, emphasized in the left SCM muscle.

CT: Inconspicuous intracranial findings.

Vertebral angiogram: Normal visualization of the vertebrobasilar circulation.

Neurological findings: Apart from the ST symptoms, there was a sensory disturbance detectable on the ulnar side side of the right forearm and the right hand. The remaining neurological findings were normal.

Operative findings: On the right side, the relationship of the of the SANR to the posterior root C1 corresponds to type IV. The anastomosis is strong developed. There is a small neuroma at the posterior root C1. There also is an anastomosis between the SANR and the posterior root C2 as well as to the anterior root C2.

On the left, the relationship between the SANR and the posterior root C1 corresponds to type III. In addition there are ananastomoses of the SANR to the posterior root C2 and to the anterior root C2. A part of the accessory root perforates the ligamentum denticulatum from dorsal to ventral and connects here with the motor C2 root. In addition, there is a neurovascular contact between the accessory nerve root and the vertebral artery.

Postoperative course: Initially, up to the time of discharge no significant change in the symptoms was recognized. Pain relief was reported. In the further course, the pain regression proved to be constant. Approximately 6 weeks after the operation, there was a gradual improvement of the

symptoms. The head now could be brought into the central position by voluntary motor control and also could be held. Driving a car became possible again. 9 months after the operation, the ST symptoms had largely disappeared, the head was straight and turning the head to the left was possible again. One year after the surgery, there was a slight recurrence of torticollis. Under intensive physiotherapy treatment, however, it was possible to achieve a satisfactory improvement and stabilization of the condition. Overall, the result now is considered to be significantly improved.

Case 20: 50 years old, female, operation October 1984

Diagnosis: Pronounced horizontal ST with head turned to the left

Medical history: 1 1/5 years before the operation there was a progressive development of horizontal ST with head turning to the left. At the same time a hypertrophy of the right SCM muscle developed. It is a clonic ST with jerky, involuntary movements of the head inrapid succession to the left. The complains began with abnormal sensationes and headache lasting several days. In the short term, under medication with Dartal an improvement of the ST symptoms could be achieved. Under stress, however, the symptoms immediately worsened again. “Geste antagoniste” with touching the chin with the left hand.

Conservative therapy: ST gymnastics, bio-feedback training, medication with Akineton, Dociton, Dartal, Xylotocan, Tia-pridex.

Diagnostics:

Neurological findings: Except for the ST symptoms with shooting innervations emphasized in the right SCM muscle the findings were regular. No indications for the presence of an extrapyramidal movement disorder.

EMG: Evidence of neurogenic damage to the nerves supplied by the accessory nerve on the right. No signs of tremor.

CT: Inconspicuous intracranial findings.

EEG: Normal alpha-EEG.

Vertebral angiogram: No evidence of pathological changes.

Operation: On the right side, the relationship of the SANR to the posterior root C1 corresponds to type IV. This anastomosis is strongly developed. In the area of the anastomosis there is a pinhead-sized small neuroma at the accessory nerve root.

On the left, the relationship of the SANR to the posterior root C1 corresponds to type III. An anastomosis of the SANR to the posterior root C2 was found. There is also a neuroma on the accessory root at the level of the foramen magnum.

On both sides, the small tumors of the SANRs could be dissected. The anastomoses of the posterior roots C1 and C2 are cut and resected. Since neurovascular contacts are not present, the padding of the accessory roots is omitted.

Histologic findings: The two thickenings on the SANRs indicate a severely degenerated part of the nerve with extensive Schwann-cell proliferation, which appears schwannoma-like. The resected posterior root C1 on the left shows a marked nerve with adjacent spinal ganglion cells without pathological changes.

Postoperative course: Initially a reduction of the impulses the right SCM muscle was observed. After completion of the intensive follow-up treatment only a slight improvement of the symptoms and a slight accessory-paresis on the left was seen. The preoperative head and neck pains had disappeared. Gradually there was a further improvement of the symptoms. The result of the treatment is considered as improved.

Case 21: 24 years old, female, surgery November 1984

Diagnosis: Horizontal ST with head turn to the right and and a minor retroflector component

Medical history: 6 months before the operation there was increasing pain in the left neck region combined with neck stiffness. This was followed by the rapid development of horizontal ST symptoms. The head could only be turned to the left with great effort. At the same time a clear hypertrophy of the left SCM muscle with hardening of the neck muscles was seen. The head misalignment appeared to be largely fixed. In the following a slight retroflection of the head was added. No significant previous illnesses. By conservative treatment no improvement in the symptoms and pain could be achieved.

Conservative therapy: physiotherapy, medication with Akineton, Dartal, Dociton, Xylotocan, Tiapridex. Under Xylotocan, side effects occurred in the form of nausea and vomiting.

Diagnostics:

Neurological findings: Apart from the ST symptoms, the neurological findings were unremarkable. There was no evidence of an extrapyramidal motor movement disorder.

EMG: On the left side, the trapezius and the SCM muscles showed increased polyphasias without evidence of recent neurogenic damage. On the right side, discrete signs of old neurogenic damage in the upper part of the trapezial muscle.

CT: No evidence of pathological changes.

X-ray: No pathological changes recognizable.

EEG: Inconspicuous brain wave curve.

Surgery: On the right side, the relationship of the SANR to the posterior root C1 corresponds to type III. The posterior

root C1 maintains an anastomosis with the accessory root and has a ganglion-like structure in the anastomosis area. The SANR is firmly attached to the vertebral artery. There is an anastomosis between the SANR and the posterior root C2.

On the left, the relationship between the SANR and posterior root C1 corresponds to type III. The SANR is anastomotically connected to the strongly developed posterior root C1. It is integrated into the dural passage of the vertebral artery and is firmly adherent there. In addition, there is an anastomosis of the accessory root to the posterior root C2.

Course: 3 weeks after the operation there was a clear improvement of the ST symptoms. 10 months after the operation tion, the patient was able to hold her head straight, turning the head to the left was possible again. The patient is back at work today. The treatment result is rated as good.

Case 22: 54 years old, male, surgery June 1985

Diagnosis: Horizontal ST with head turned to the right

Medical history: 7 years before the operation, the patient developed of a horizontal ST with head turn to the right. Temporarily remission of the symptoms under conservative treatment. 3 years before the operation recurrence of the ST symptoms, now associated with severe neck pain. 1 year before the BMLA, a highly cervical epidural spinal cord stimulation was performed, but this did not had any effect on the ST symptoms. On the contrary, the involuntary head misalignment increased. The symptoms appeared as a fixed head turn to the right with an anteflectory component. At the same time development of hypertrophy of the left SCM muscle without any significant innervations.

Conservative therapy: ST gymnastics, biofeedback, psycho therapy, spinal cord stimulation, medication with Akineton, Dartal.

Diagnostics:

Neurological findings: Apart from the ST symptoms, unremarkable neurological findings without evidence of a central movement disorder.

EMG: Increased polyphasic potentials in the SCM muscle on both sides without reliable evidence of neurogenic damage.

EEG: Normal alpha EEG.

CT: Inconspicuous intracranial findings.

Surgery: On the right side, the relationship of the SANR to the posterior root C1 corresponds to type IV. In the anastomosis area there is a pinhead sized nodule which is heavily vascularized.

On the left side, also the relationship of the SANR to the posterior root C1 corresponds to type IV. The SANR is drawn into the intervertebral foramen C1 and is firmly attached to the vertebral artery. In addition, there is an anastomosis of the SANR with the posterior root C2.

Histologic findings: The sections contain parts of a spinal ganglion and a longitudinally impacted nerve root. In addition, there is arachnoid connective tissue that is firmly connected to the nerve root. The findings suggest a scarred, healed inflammatory process.

Postoperative course: Already 1 week after the operation the symptoms and the pain were clearly improved. 9 months after the operation almost complete regression of the ST symptoms. The patient is now symptom-free and can go about his work. The outcome can be described as very good.

Case 23: 49 years old, female, surgery October 1980

Diagnosis: Rotatory ST with tilting of the head to the right

Medical history: 3 years before the operation, initially only lying down, later increasingly also when sitting, standing and walking, involuntary tilting of the head with inclination of the head to the right occurred. A psychogenic trigger was suspected and psychotherapy was suggested. However, this could not influence the clinical picture. On the contrary, there was an intensification of the symptoms, so that the right ear almost touched the raised shoulder. All other conservative measures remained unsuccessful.

Conservative therapy: psychotherapy, medication with Tranxilium, Tiapridex, Akineton

Diagnostics:

Neurological findings: pronounced rotatory ST with head tilt to the right. Significant hypertrophy of the left SCM muscle.

EMG: The findings suggest a neurogenic lesion of the left of the neck muscles supplied by the left accessory nerve.

CT: No evidence of pathological changes.

Vertebral angiogram: The left PICA forms a cavity below the level of the occipital foramen. Otherwise inconspicuous visualization of the vertebraobasilar area.

Operation: On the right side, the relationship of the SANR to the posterior root C1 corresponds to type IV. The SANR is pulled into the intervertebral foramen C1 and is adherent to the vertebral artery here.

On the left, the relationship of the SANR to the posterior root C1 corresponds to type IV. Due to the short poterior root C1, the SANR is pressed onto the vertebral artery.

Postoperative course: 6 months after the operation there was

a gradual gradual regression of the ST symptoms. The patient now was able to hold her head straight again for a longer time and to turn the head to both sides. The treatment result is seen by the patient as improved, objectively as good.

Case 24: 41 years old, male, surgery March 1981

Diagnosis: Pronounced rotatory ST with head tilt to the right

Medical history: 8 years before the operation the patient developed symptoms with head inclination to the right, beginning with a feeling of tension in the area of the right right shoulder blade, which lasted for several months. 3 months after the first symptoms the head began to wobble, which slowly subsided as the head tilt increased. Finally, despite various conservative conservative treatment attempts, the symptoms were largely fixed. With the onset of the disease there was an alcohol abuse, as the patient felt a relief of symptoms under the influence of alcohol. The patient attempted suicide one year before the operation.

Conservative therapy: psychotherapy and medication with Decentan and Tiapridex.

Neurological findings: Apart from the ST symptoms with a hypertrophy of both SCM muscles emphasized on the left, there is a sensory disturbance in the right forearm and the right hand corresponding to dermatome C8. The remaining neurological findings are normal.

EMG: More pronounced in the right SCM muscle than on the left, less also in both trapezius muscles, there are partly old, partly also more recent signs of neurogenic damage.

CT: Pathological changes are not recognizable.

Vertebral angiography: Arcuate mediocaudal course of the left PICA. Other pathologic changes are not recognizable.

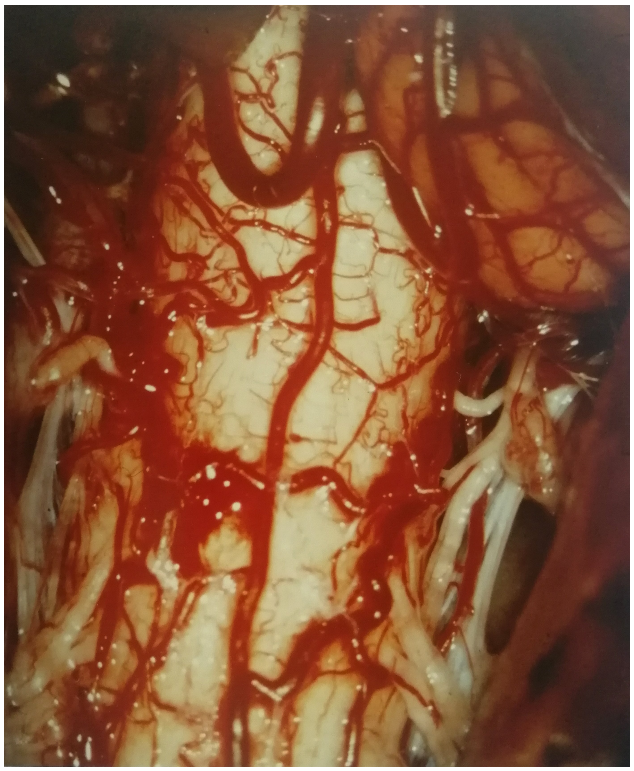
X-ray: Right curvature of the cervical spine with increased kyphosis in the cervicothoracic transition and corresponding lordosis in the cervicooccipital junction. Spondylosis deformans at the level of CV5, 6 and 7.

EEG: Relatively slow alpha EEG with questionable slight allcommon change.

Operative findings: On the right side, the relationship of the SANR to the posterior root C1 corresponds to type IV. The posterior root C1 has a ganglion-like formation near the anastomosis. At the level of the dura entry of the vertebral artery, a posterior spinal artery arises, which crosses under the SANR. A loop of the PICA, which runs caudal in a large arc has a neurovascular contact with cranial parts of the accessory nerve root.

On the left side, the relationship of the SANR to the posterior root C1 also corresponds to type IV. There is a close vascular contact between the SANR to a strong spinal artery that departs directly from the dural passage of the vertebral artery (Fig. 13).

Fig. 13: Case 24, Surgical site



Postoperative course: Three months after the operation improvement in ST symptoms with good control of head mobility. The head could now also be turned to the left. However, the head was still clearly tilted when concentration was distracted. 1 year after the operation, the hypertrophy of the SCM muscles had regressed. At rest, especially when sitting, the head can be held completely straight. However, the head is still clearly tilted when walking. 3 1/2 years after the operation, the patient died as a result of suicide. By this time, the ST symptoms could be assessed as improved.

Case 25: 44 years old, male, surgery September 1985

Diagnosis: Pronounced horizontal ST with head turn and and slight head tilt to the right

Medical history: 12 years before the operation the patient developed ST, initially with head tilt to the left. After temporary spontaneous remission of the clinical picture, a recurrence occurred with head tilt to the right. In addition a shoulder-arm syndrome developed on the right with pain radiating to the dorsal side of the right upper arm. All conservative treatment attempts were unsuccessful.

Conservative therapy: Diazepam, Tiapridex, Akineton

Diagnostics:

Neurological findings: Apart from the ST symptoms with a pronounced head turn and slight head rotation to the right, the neurological findings were normal. There were no signs of torsion dystonia or any other extrapyramidal disorder.

EMG: Neurogenic damage in the left SCM muscle.

CT: Inconspicuous intracranial findings.

X-ray: Significant malposition of the cervical spine with deflection to the right.
Surgery: On the right, the relationship of the SANR to the posterior root C1 corresponds to type IV. In the region of the anastomosis between the SANR and the posterior root C1 there is a pinhead-sized structure, which is heavily vascularized.

On the left side, the relationship between the accessory nerve to the posterior root C1 corresponds to type IV. The SANR is drawn into the intervertebral foramen C1. In addition, there is a further anastomosis to the posterior root C2. Neurovascular contacts were not found apart from the adhesion of the left accessory root with the vertebral artery at the dural passage.

Postoperative course: Immediately after the procedure the head constraint was relaxed. The right-sided brachialgia had disappeared. Shooting innervations were no longer recognizable. 10 months after the operation gradual regression of the torticollis symptoms occurred. 18 months after the operation, the patient's condition can be rated as improved. When sitting, the head can be kept straight for a long time. When walking, control is of the head is still considerably more difficult. The patient stated that he is able to live with the existing residual symptoms.

Case 26: 45 years old, male, surgery October 1985

Diagnosis: Horizontal ST with head turned to the right

Medical history: 22 years before the operation, following a head trauma to the skull had gradually led to the development of an involuntary head posture. Before the operation the symptoms presented as a pronounced ST with a head turn to the right. All conservative treatment measures remained unsuccessful.

Conservative therapy: Limbatril, Muscle Trancopal, Tiapridex, Akineton
Diagnostics:

Neurological findings: Purely horizontal ST with turning of the head to the right and hypertrophy of the left SCM muscle. Shoulder elevation on the right. The remaining neurological findings are normal.

EMG: Emphasized in the left SCM muscle there are clues, which can be interpreted as neurogenic damage to the muscle.

X-ray: Incomplete closure of the vertebral arch from CV3 to CV5. Spondylarthrosis in CV2 on the right and right accentuated arthritis of the atlanto-occipital joints.

CT: Large calcifications of the choroid plexus of both lateral ventricles. Otherwise unremarkable findings.

Operative findings: On the right side the findings correspond to type I. The posterior root C1 is not present. The posterior root C2 is anastomotically connected to the SANR. Otherwise, the right SANR is completely unobstructed.

On the left, the relationship of the SANR to the posterior root C1 corresponds to type IV. There is also an asnastomosis to the posterior root C2. In the area of the anastomosis between the accessory root and the posterior root C1 there is a vessel-injected distension of the SANR. It is in contact with the vertebral artery and is adherent to it in the area of its dura passage.

Postoperative course: Immediately postoperatively, the malposition of the head was considerably improved. The turn of the head to the left was possible without effort. 17 months after the operation, the treatment result could be assessed as good.

Case 27: 45 years, female, surgery March 1986

Diagnosis: Horizontal ST to the right

Medical history: 3 years before the operation an involuntary head tilted to the right. At the same time, the left SCM muscle hypertrophied. When standing and walking, there were jerky turning movements of the head to the right. When lying down, however, the head was still. When tense and nervous, the symptoms increased. 2 years before the operation neck pain radiating into both shoulders, at times also into the upper arms occurred. Conservative treatment measures were unsuccessful.

Conservative therapy: bio-feedback training, Dartal, Xylo-tocan, Diacepam

Diagnostics:

Neurological findings: Apart from the ST symptoms with clonic clonic movement restlessness of the head and hypertrophy of the left SCM muscle, the neurological findings were normal.

EMG: The findings suggested predominantly old neurogenic damage in the left SCM and left trapezial muscles.

Vertebral angiography: Inconspicuous visualization of the vertebral-basillar stromal area.

CT: Inconspicuous intracranial findings.

EEG: Intermittent decelerations, but without definite focal findings and without signs of a specific increase in excitability.

Operation: On both sides the relationship of the SANRs to the posterior roots C1 corresponds to type I. On the right side, the SANR runs taut over the PICA arising at the dural passage of the vertebral artery. A very irritable, fine nerve branch of the SANR runs in the direction of the first attachment of

the denticular ligament. Whether this is possibly a very fine posterior C1 root cannot be determined.

On the left side, the SANR maintains vascular contact with a caudally directed PICA loop.

Postoperative course: Immediately postoperatively, the patient initially reported a significant improvement in her complaints. The clonic innervations occurred only occasionally. At the time of discharge, however, an increase in symptoms was recognizable. 12 months after surgery, there was a clear regression of ST symptoms. The treatment result is rated as good.

Case 28: 45 years old, male, surgery May 1986

Diagnosis: Combined rotatory-horizontal ST with head inclination to the right and turning of the head to the left

Medical history: 3 years before the operation, the patient developed symptoms of torticollis with tilting of the head to the right and slight turn to the left. Moderate hypertrophy of the right SCM muscle. At the same time shoulder elevation on the right. All conservative treatment measures were unsuccessful.

Conservative therapy: psychotherapy, ST gymnastics, Diaze-pam, Dartal, Melasil

Diagnostics:

Neurological findings: Apart from the torticollis symptoms the neurological findings were normal, in particular there were no indications of the presence of a central motor disease.

EMG: There are indications of an old neurogenic damage of the muscels supplied by the right accessory nerve.

CT: Inconspicuous intracranial findings.

Vertebral angiogram: Very deep origin of the left PICA with loop formation far below the level of the foramen magnum. The right PICA also has a far caudal extending loop.

EEG: Unobtrusive brain wave pattern.

Surgical findings: On the right, the relationship of the SANR to the posterior root C1 corresponds to type I. Notable particularities are not found.

On the left side, the relationship of the SANR to the posterior root C1 corresponds to type III. The left PICA, which forms a caudal corkscrew-like loop, is firmly adherent to the SANR, which is taut over the vertebral artery and the upper tooth of the dentate ligament.

Postoperative course: At the time of discharge no change of ST symptoms. Compared to the preoperative findings, a paresis of the left trapezial muscle with impaired lateral arm elevation on the left was noted. 9 months after the operation an extensive regression of the left-sided accessory nerve paresis occurred. The ST symptoms appeared only slightly improved at this time.

Case 29: 48 years old, female, surgery October 1986

Diagnosis: Severely pronounced combined rotatory-horizontal ST with tilting of the head to the left and turning of the head to the right. Condition after stereotactic thalamo- and subthalamotomy on the right, 4 years before BMLA. Basilar impression.

Medical history: 8 years before the BMLA, there were initially occasional myoclonic convulsions with tilting of the head to the left shoulder. Within 2 years a pronounced clonic torticollis, initially with tilting of the head to the left and minor retrocollis symptoms occurred. Painful tension

developed in the left upper part of the trapezius and the left splenius capitis muscles. The ever increasing painful tension of the shoulder and neck muscles, particularly on the left, required the intake of analgesics in considerable doses and the daily injection of local anaesthetics into the tense muscles. In addition to the torticollis symptoms, a considerable scoliosis developed at the same time with a bending of the upper body in the hip to the left. There was also an atlas hypoplasia with dens elevation and simultaneous basilar impression. Due to the diagnosis of “torsion dystonia", the indication for thalamo- and subthalamotomy on the right was made. The procedure was performed 4 years before BMLA. Immediately postoperatively, the torticollis symptoms had initially disappeared completely. However, after the coagulation edema subsided, the symptoms partially reappeared. For 6 months a moderate improvement in the symptoms was observed. Then again the development of a high degree of head obliquity occurred, which, however, had changed due to the previous stereotactic procedure. Pronounced torticollis symptoms developed with a head tilt to the left, head turned to the right and at the same time very strong retrocollis symptoms. A variety of conservative treatment measures and the severing of the left SCM muscle also did not lead to any improvement.

Conservative therapy: Sarotene, Tavor, Akineton, Normabrain

Diagnostics:

Neurological findings: High degree of head tilt to the left, head turn to the right and retrocollis. Hypertrophy of the trapecius and splenius capitis muscles on the left. Shoulder elevation on the left with fencing position. Torso inclination to the right. The remaining neurological findings were normal, in particular there were no indications of the presence of torsion dystonia. Motor dexterity and fine motor skills were normal on both sides.

EMG: Evidence of minor neurogenic damage in the right muscles supplied by the right accessory nerve.

CT: Intracranial findings unremarkable.
EEG: Unremarkable brain wave pattern.

X-ray: Complex malformation in the area of the craniocervical passage with basilar impression and block vertebrae formation CV2/3.

Vertebral angiogram: Deep PICA loop reaching below the level of the foramen magnum on the left.

Surgery: As stereotactic surgery had already been unsuccessful, the diagnosis of a ST is assumed, as dysplastic changes in the craniocervical junction, which may be the cause of irritation of the SANRs or the upper cervical spinal nerves.
On the right side, the relationship between the SANR and posterior root C1 corresponds to type IV. The SANR is closely attached to the foramen of the root C1 and is in close contact with a posterior spinal artery that arises directly from the vertebral artery. In addition, a vascular bundle of this artery is wound around the SANR.

On the left, the relationship of the SANR to the posterior root C1 corresponds to type I. However, there is an anastomosis of the SANR to the posterior root C2. Otherwise, the SANR runs unobstructed without neurocascular contacts.

Postoperative course: In the first few days postoperatively symptoms were unchanged. After the wound pain had subsided, there was a significant reduction of symptoms with reference to the preoperative severe left-sided neck and shoulder pain. In the further course the torticollis symptoms improved within 6 months. Even 1 year after the operation the torticollis symptoms were still clearly improved, although there was still a pronounced retrocollis symptomatology, which, however, was due to the previous severing of the SCM muscle. In the further course the complex spinal misalignment then led to the development of C8 root irritation symptoms

with severe pain. Finally, there was also a further increase in the head misalignment, in particular clonic innervations, so that 60 months after the operation, the overall condition was unchanged compared to the preoperative condition. 4 years after the operation there were indications for the presence of a segmental torsion dystonia, as oromandibular dyskinesia could now sometimes be observed.

Case 30: 54 years old, female, surgery November 1982

Diagnosis: Pronounced rotatory horizontal ST with tilting and turning of the head to the left

Medical history: 8 years before the BMLA there was a gradual and painless increasing involuntary tilting of the head with turning to the left. In the following years, pain complaints developed increasingly with neck and back pain. Despite various attempts at conservative treatment, the disease progressed over the years. 3 years before the BMLA a stereotactic thalamotomy was performed on the right, which however, did not result in any improvement of the symptoms. 1 year before the BMLA, a myotomy of the left SCM muscle was performed. This also did not influence the symptoms. While the torticollis symptoms were less pronounced when lying down, a permanent contraction of the left SCM muscle, and to a lesser extent also of the right trapecius and splenius capitis muscles occurred when walking. Occasionally, shooting dyskinesia in the area of the left shoulder was observed.

Conservative treatment: Attempts at drug treatment with over 30 medications, including Valium, Akineton and Dartal, Bio-feedback treatment, acupuncture, neural therapy and physiotherapy.

Diagnostics:

Neurological findings: Right convex cervical spine scoliosis with left shoulder elevation, the head is very strongly

tilted and turned to the left. The voluntary motor movement of the head to the right is not possible. The remaining neurological findings are normal.

EMG: The findings are consistent with old neurogenic damage in the left SCM muscle and for more recent neurogenic damage in the right SCM muscle.

CT: Inconspicuous intracranial findings.

X-ray: Pronounced spondylotic changes at the level of CV4/5 and CV5/6 with right convex scoliosis.

Vertebral angiogram: The left PICA forms a loop reaching below the level of the foramen magnum with an otherwise unremarkable presentation of the vascular system.

EEG: Beta-EEG without evidence of pathological changes.

Operation: On the right side, the relationship of the SANR to the posterior root C1 corresponds to type IV. A further anastomosis is found to the posterior root C2. There are no vascular-nerve contacts.

On the left, a posterior root C1 corresponding to type I is not present. The SANR has a very tense course. At the level of the foramen magnum there is an adhesion of the SANR with the deep PICA loop. In the area of this neurovascular contact the SANR appears altered by connective tissue.

Postoperative course: After the procedure, initially there was seen an improvement of the torticollis symptoms. The patient was able to keep her head straight, albeit only for a short time. However, there was no significant improvement in the further course and the result of the treatment was regarded by the patient herself as unimproved.

Case 31: 41 years old, male, surgery February 1983

Diagnosis: Combined rotatory-horizontal torticollis with tilting and turning of the head to the left
5 years before the operation, increasingly an involuntary turning of the head to the left occurred. The intensity of the symptoms remained fluctuating until 1 year before the operation. All conservative measures were unsuccessful. Temporarily, treatment with Tocainide-HCl resulted in an improvement. Overall, however the course of the disease remained progressive. In addition to the torticollis symptoms, the patient had a history of mesenteric and splenic vein thrombosis 10 and 1 year before the operation. Thrombophlebitis migrans is discussed as the cause. Marcumar treatment was initiated.

Conservative therapy: Bio-feedback training, Tiapridex, Dartal, Xylotocan, Valium

Diagnostics:

Neurological findings: Apart from the clonic torticollis symptoms with jerky head turns and simultaneous tilt of the head to the left, the neurological findings are normal. The right SCM muscle is hypertrophied. The torticollis symptoms are more pronounced at rest than during exertion. Pain is not complained of.

EMG: Neurogenic damage in the muscles supplied by the accessory nerve, pronounced on the right.

Vertebral angiogram: Deep PICA loop right below the level of the foramen magnum

CT: No evidence of pathological changes intracranially.

Surgery: On the right side, the relationship of the SANR to the posterior root C1 corresponds to type IV. In addition, there is a pronounced neurovascular contact of SANR with the abnormally deep PICA loop, which accompanies the nerve over a distance of approx. 2 cm. The PICA is firmly adherent to the SANR.

On the left, corresponding to type I, there is no posterior root C1. The accessory nerve is completely unobstructed here. Postoperative course: After initially good regression of the ST symptoms and reintegration into the work process 14 months after the operation there was a renewed deterioration again. There were now severe tensions in the right trapezius muscle with severe neck pain. The hypertrophy of the right SCM muscle had regressed compared to the preoperative state. 2 years after the operation, the ST symptoms corresponded to the preoperative findings. The treatment result is to be regarded as unimproved.

Case 32: 30 years old, male, surgery September 1983

Diagnosis: Pronounced combined rotatory-horizontal ST with turning and tilting of the head to the left

Medical history: 1 year before the operation the patient noticed an increasing neck stiffness. This was followed within a few days by involuntary turning movements of the head to the left with simultaneous rotatory component. All conservative treatment measures did not lead to any improvement in the progressive torticollis symptoms. In addition to acupuncture, scalenus blockades and psychosomatic treatment, transcutaneous and epidural DCS was performed. 1/2 year before the BMLA, high cervical epidural electrodes for spinal cord stimulation were implanted. Despite correct electrode position no stimulation success could be achieved.

Conservative treatment: Akineton, Dartal

Diagnostics:

Neurological findings: Pronounced ST with turning and lateral tilt of the head to the left. While the head can still be held straight in the short term, there are often innervations with jerky head inclination. Hypertrophy of the SCM muscles is not recognizable. Otherwise the neurological status is normal.

EMG: The findings suggest minor, recent neurogenic damage in the right SCM muscle with unobtrusive findings on the left. On the left side there is a tremor with frequency of 8/sec.

CT: Intracranial findings unremarkable.

Vertebral angiogram: Inconspicuous visualization of the vertebral-basilar artery area.

Surgery: On the right side, the findings correspond to type III. There is a close anastomotic connection between the posterior root C1 to the SANR, which is adherent to the right vertebral artery.

On the left, the relationship of the SANR to the posterior root C1 corresponds to type IV. Because of the anastomosis with the posterior root C1, the SANR is drawn into the C1 foramen. A further anastomosis is found to the posterior root C2. There is also close contact between of the accessory root to the posterior spinal artery, which connects the SANR with a vascular bundle. A few mm above the junction with the vertebral artery, the SANR is surrounded by a peasized arachnoid cyst in the shape of a cuff.

Postoperative course: An improvement of the torticollis symptoms was not observed. On the contrary, the symptoms were progressive, so that now presence of a torsion dystonia could be assumed. In addition to the symptoms of torticollis symptoms, there were now oromandibular dyskinesias and restlessness of the left hand.

Case 33: 51 years old, male, surgery November 1983

Diagnosis: Combined rotatory-horizontal ST with head tilt to the left and head turn to the right

Medical history: Since 11 years before the operation the patient suffered from recurrent cervical spine complaints.

6 years before the operation, he initially noticed a slight involuntary pulling of the head to the right when lying down. In the following period the symptoms progressed. Finally, a pronounced ST developed with a strong inclination of the head to the left and turning to the right. In addition, the patient complained of a painful left shoulder-arm syndrome with a sensory disturbance corresponding to dermatome C7 on the left. All conservative treatment measures were unsuccessful, so that 2 years before the BMLA an attempt with cervical epidural electrical stimulation of the cervical medulla was carried out. After an initial improvement, however, the symptoms quickly returned in old form.

Conservative treatment: Valium, Lexotanil, Akineton, Dartal

Diagnostics:

Neurological findings: In addition to the ST symptoms, there is a root compression syndrome C7 left with loss of TSR and a sensory disturbance corresponding to C7. No indications for torsion dystonia or another extrapyramidal disease.

EMG: In the SCM muscle on the right and in the trapezial muscle on the left are signs of neurogenic damage in the form of clearly enlarged and widened potentials.

CT: No intracranial abnormalities.

Vertebral angiogram: Inconspicuous visualization of the vertebral-basilar artery area.

Surgery: On the right side, the relationship of the SANR to the posterior root C1 corresponds to type IV. As a result of the anastomosis to the posterior root C1, the SANR is attached to the PICA leaving the vertebral artery through the dural passage and is adherent.

On the left, the relationship of the SANR to the posterior root C1 corresponds to type III. The SANR is pulled into the foramen C1 and fixed on the vertebral artery. The posterior

root C1 is bulbously distended, clearly vascularized and yellowish in color. It imponates like a spinal ganglion. There is also an anastomosis of the accessory root to the posterior root C2.

Postoperative course: Shortly after surgery improvement and loosening of the head posture was already noticed. The left-sided C7 root compression syndrome regressed. The patient was painfree. In the further course, the condition stabilized, the torticollis symptoms were considered to have improved. However, the patient himself was not satisfied with the result of the treatment and assessed his condition as condition as unimproved.

Case 34: 41 years old, female, surgery May 1983

Diagnosis: High grade clonic antecollis with rotation of the head to the right

Medical history: 21 months before the operation there was an uninvoluntary head tilt to the right. The symptoms occurred acutely and showed rapid progression. In the following the patient was treated in psychiatric clinics for 9 months without success. After initially predominantly tonic symptoms were followed by cloniform twitching of the head, which also affected the upper body. Six months before the operation there was a significant, 4-week lasting, remission of symptoms. Finally the symptoms developed again in full strength. In the foreground of the complaints severe pain now was constant, that moved from the neck to the back of the head. At times, the patient also suffered from bilateral shoulder-arm pain. Intermittently numbness in the dermatome C8 on the left side occurred. Occasionally there were violent shaking movements of the head tilted to the right in anteflection. For this reason, the patient usually held her head with both hands. Multiple attempts at treatment with medication were without success.

Conservative therapy: Psychotherapy, Tiapridex, Dartal,

Akineton

Diagnostics:

Neurological findings: Extreme torticollis symptoms with antecollis and tilting of the head to the right as well as clonic shaking movements of the head. In addition there was also a sensory disturbance corresponding to dermatoma C8 on the left. The other neurological findings were normal.

EMG: In the muscles supplied on both sides by the accessory nerve pathologically deformed action potentials were found, which were indicative of neurogenic damage.

CT: Intracranial findings unremarkable.

Vertebral angiogram: The left PICA has a deep outlet from the vertebral artery with deep loop formation reaching below the level of the foramen magnum. There also is a wide PICA loop on the right.

Operation: On the right side the relation of the SANR to the posterior root C1 corresponds to type IV. The posterior root C1 enters the spinal canal next to the vertebral artery into the spinal canal and has a ganglion-like structure, which connects to the SANR. The SANR is firmly attached to the vertebral artery. A further anastomosis to the posterior root C2 is found.

On the left, the relationship of the SANR to the posterior root C1 also corresponds to type IV. The PICA, arise from the dural passage of the vertebral artery, forms a loop extending far caudally and to the right. Both SANRs have contact with this vessel. In addition, the accessory root is stretched over the PICA close to its origin from the vertebral artery.

Postoperative course: After initial improvement of the symptoms, a renewed significant deterioration was noted in the long term. The violent percussive movements of the head to the right increased. The left SCM muscle became

increasingly hypertrophied. The gait was unsteady and unsure. In the further course of the disease symptoms spread to the right arm and upper body. Attempts at drug treatment with Dartal and Xylotocan did not affect the symptoms. Biofeedback training was also unsuccessful. As the clinical picture in this patient undoubtedly developed into DT (dystonic torticollis), 3 years after the BMLA, the decision was made to perform a bilateral a stereotactic thalamo- and pallidotomy. This resulted in a clearly improvement of the massive symptoms.

Case 35: 37 years old, male, surgery March 1980

Diagnosis: ST with retrocollis and right head turn

Medical history: 10 years before the operation occasional head turning movements to the right occurred. At the same time, pain radiating from the neck pain into the left shoulder and left arm occurred. 3 years before the operation, the symptoms worsened and developed a massive retrocollis with simultaneous turning of the head turned to the right. The head shows a constant clonic restlessness of movement. The left SCM muscle is clearly hypertrophied. Only with great effort and pain it was possible to bring the head into middle position. Increasingly, in addition to the torticollis symptoms, a left shoulder-arm syndrome developed. To alleviate the symptoms, the patient useds the “Geste antagoniste“. He touches his chin with his right hand. The symptoms are roughly the same when lying down, standing and walking. In addition to the torticollis symptomatology, there was a striking lively facial expressions with pulling down the right corner of the mouth and wide opening of the eyes. All conservative treatment attempts were unsuccessful.

Therapy: bio-feedback training, Tiapridex, Haldol, Dartal

Diagnostics:

Neurological findings: Combined torticollis symptoms with retrocollis and head turning to the right. Pronounced hypertrophy of the left SCM muscle. The other neurological findings are normal. Apart from the mimic motor activity, there are no indications for the presence of an extrapyramidal motor disorder recognizable.

EMG: Massive permanent innervation in the left SCM muscle, less also in the splenius capitis and trapecius muscles on the right. No definite indication of neurogenic damage to the muscles supplied by the accessory nerve.

CT: Minor enlargement of the insular cisterns emphasized on the left with otherwise unremarkable intracranial findings.

Vertebral angiogram: Unpaired left vertebral artery.

Fig. 14: Case 35, Surgical sketch, left side
Operation: Exposure of the SANR on the left. The relationship of the SANR to the posterior root corresponds to type III. The posterior root C1 is connected to the with the SANR via a ganglion-like structure. In addition, there is an adhesion with the ligamentum denticulatum (Fig. 14). The SANR was severed when attempting to detach the ganglion from it.

Histologic findings: The resected nerve portion corresponds to a small neuroma with a severely degenerated nerve.

Postoperative course: Postoperative initially significant improvement of the torticollis symptoms with simultaneous complete accessory nerve palsy on the left. The patient was able to hold his head straight for long periods of time. 5 years after the operation, however, there are still retrocollis symptoms with spasmodic tensions of the chewing and tongue muscles. 4 1/2 years after the operation during an extreme tenseness of the chewing and tongue, a fracture occurred of 4 mandibular incisors. Because of the very clear oromandibular dyskinesia, the presence of segmental dystonia now must be assumed. The torticollis symptoms nevertheless can be judged as improved, which also corresponds to the patient's opinion. The complete accessory palsy on the left with atrophy of the suprascapular muscula and a slight scapula alata exists unchanged.

Case 36: 55 years old, female, surgery November 1980

Diagnosis: ST with retrocollis and horizontal component to the right

Medical history: 4 years before the operation there was an involuntary turning of the head to the right. 3 years later a progressive deterioration with retrocollis symptoms occurred. Hypertrophy of the left SCM muscle developed. 1/2 year before the operation the retrocollis symptoms came to the fore. In addition severe neck pain occurred, partly radiating into the left side of the face. The patient also reported intermittent

tension in her left leg. All conservative measures were unsuccessful.

Conservative treatment: Massages and talk therapy, baths, Akineton, Dartal, Tiapridex, Liorisal

Diagnostics:

Neurological findings: Fixed retrocollis symptoms with turning of the head to the right. Turning the head to the left is hardly possible. Hypertrophy of the left SCM muscle. The remaining neurological findings are normal, in particular there are no indications for the presence of an extrapyramidal motor disorder.

EMG: The findings suggest neurogenic damage to the SCM muscle, on the right more clearly than on the left. Less also pronounced in both trapezius muscle. The damage is partly fresh, partly of an older nature. In addition, a pseudomyotonic series is described.

CT: Intracranial pathological findings are not detectable.

X-ray: Pronounced lordosis of the cervical spine.

Vertebral angiogram: Inconspicuous visualization of the vertebral-basilar artery area.

EEG: With low voltage development there are no reliable lateral indications.

Operation: The relationship of the SANRs to the posterior roots C1 corresponds to type IV on both sides.

On the right side there is a vascular contact of the accessory nerve root with a posterior spinal artery at the dura-passage of the vertebral artery. In addition there is also an adhesion to the attachment of the dentate ligament. The nerve is fixed here and tortured 360 degrees.

On the left side the posterior C1 root in the area of the anastomosis has a strongly vascularized ganglion. The SANR is stretched over the vertebral artery and is adherent with this. A further anastomosis exists to the posterior root C2.

Histologic findings: The examination of the resected right-sided posterior root C1 shows a moderately pronounced Waller's degeneration with pathological Schwann-cell proliferation. The change represents the morphological substrate of chronic nerve irritation.

Postoperative course: Immediately after the operation a slight reduction in the tone of the muscles involved in the symptoms of torticollis occurred. In the further course only a slight improvement of the torticollis symptoms. Turning of the head to the right decreased and the head could be held in the middle position for a longer period of time. On the other hand, there was an increase of the retrocollis symptoms. Overall, the patient's condition can be regarded as slightly improved. The patient herself considers her condition to be unimproved.

Case 37: 43 years old, male, surgery July 1982, May 1983

Diagnosis: Combined horizontal-rotatory ST with head turn to the right and slight retroflexion

History: The disease began 3 years before our first operation and 14 days after a smallpox vaccination with a slight pull of the head to the right. In the further slowly progressive course, a combined horicontal-rotatory ST symptoms with turning and tilting of the head to the right developped. These were intermittent occurring, skidding movements. At the same time hypertrophized the upper part of the left trapezius muscle. Finally, there was also a shoulder-arm syndrome on the right. Up to this point the other neurological findings had been unremarkable. Two years after the onset of the symptoms a left subthalamotomy was performed. Postoperatively the torticollis symptoms remained essentially unchanged.

However, a side effect of the procedure was a slight right hemiparesis with impaired fine motor function of the right hand. In addition, a speech disorder and a reactive depressive mood was noticeabel.

Conservative therapy: psychosomatic treatment, physical therapy, medication with Tiapridex, Valoron, Dartal, Muskeltrancopal

Diagnostics:

Neurological findings: ST with turning of the head to the right, inclination to the left and slight retrocollis symptoms. Hypertrophy of the splenius muscle on the left. Arm accentuated hemi-hypotonia and hemi-ataxia on the right, intention tremor on the right, mild dysarthria. No evidence of torsion dystonia.

EMG: Increase in the potential amplitudes on derivation from the SCM muscels (right 7.3 mV, left 6.2 mV). No definite neurogenic damage detectable in the examined muscles.

CT: Except for the stereotactic coagulation defect with a diameter of 5 mm in diameter in the left subthalamic region, there are no no intracranial pathologic findings.

X-ray: Left convex malposition with spondylarthrotic and spondylotic changes in CV5, 6 and 7.

Vertebral angiogram: Inconspicuous visualization of the vertebrobasilar artery area.

EEG: Regular alpha-wave basic pattern.

Operation: 8 months after the stereotactic procedure, the BMLA was performed.

On the right side an anastomosis was found between the SANR and the posterior root C1 which corresponds to type IV.

On the left side, there were also anastomoses between the SANR and the posterior roots C1 and C2, which also coresponds type IV. A caliber-strong PICA, which after its exit from the vertebral artery 5 mm parallel and in direct contact with the SANR runs downward until shortly before its passage into the jugular foramen. At the level of the foramen magnum, a branch of the PICA is also in contact with some fine accessory nerve roots, which join with the SANR at the level of the vertebral artery entry.

Postoperative course: Initially complicated by a massive abacterial meningitis. At the final examination an improvement of the torticollis symptoms was noted. In the course of the following year, with pre-existing spondylotic changes in the cervical spine, a C4 root syndrome developed on the left with significant pain radiating into the arm.

On re-presentation, the ST symptoms appeared to have improved, but by no means regressed. Therefore, 10 months after the exposure of the craniocervical junction, this area was revised: Significant achnoidal adhesions were found, which were coming from caudal up to the C1 could be removed. Above this area, the dura could no longer be separatet from the spinal cord. The left SANR was explored and severed. Likewise the left ventral C2 root and the dorsal C1 root also were severed. The right ventral C2 root was rarified. As a further procedure was not possible due to the adhesions, the procedure was terminated.

Postoperatively, the patient initially recovered slowly, but could eventually be fully mobilized. Two weeks after surgery, a hydrocephalus internus developed, which was drained. However, the patient's condition continued to deteriorate with signs of increasing respiratory insufficiency. Tracheotomy and mechanical ventilation became necessary. Finally, after the development of pneumonia, pulmonary fibrosis developed with a corresponding diffusion disorder. Death occurred three months after the intervention. A necropsy was performed and the findings were described in the chapter on the previously published neuropathological findings published to date (see p. 29).