Quality of life after decompressive craniectomy for malignant middle cerebral artery infarction

Malignant middle cerebral artery (MCA) infarction is a devastating condition leading to early death in nearly 80% of cases due to the rapid rise of intracranial pressure despite maximum medical management of the ischaemic brain oedema. Decompressive craniectomy (DC) has been proposed to prevent brain herniation in malignant MCA infarction, but it remains controversial in the absence of randomised controlled trials and because of the fear of a severe residual disability after surgery. We present herein the results of a quality of life assessment using patient and proxy versions of the Stroke Impact Scale (SIS) in eight patients 12–30 months after craniectomy for malignant MCA infarcts.

Methods

Between March 1999 and November 2000, all consecutive patients with malignant MCA infarction were treated by DC and durotomy for malignant MCA infarcts.

Ten patients were included (eight men and two women, mean (SD) age 41 (12) years, range 15–54). The mean (SD) NIH score scale at admission was 21 (3), range 16–25. Five patients had a left sided stroke with severe aphasia. The mean time between stroke onset and surgery was 65 (68) h, range 12–252. One patient had a late DC because of recurrent MCA infarct at day 9 after the first stroke. All patients had signs of temporal brain herniation before surgery including uni- or bilateral mydriasis (9/10), Cheynes-Stokes hypoventilation (8/10), or decerebration (6/10). The mean (SD) duration of hospitalisation in the intensive care unit was 22 (20) days, range 3–58. Two patients died, one from a cerebral abscess and the other from a large epidural hematoma.

All living patients (8/10) were followed for a mean (SD) duration of 21 (21) months, range 12–30. All were managed in a specialised stroke rehabilitation unit with a mean (SD) hospital stay of 12 (11) months, range 4–24, after which they returned home with either home rehabilitation facility or day hospital care. At the end of follow up, 7/8 patients had an mRS ≤ 4 (table 1). The mean (SD) NIH stroke score was 13 (4), range 8–18.

The two youngest patients had the best scores on disability (mRS = 2) and were fully independent for the activities of daily living (BI = 90) (table 1).

The 64 SIS items could be measured in all patients except patient 7 who had severe aphasia (table 1). The proxy version of the SIS was administered to a close relative (five spouses, two parents) or an employed caregiver (one). The mean (SD) patient assessment of global perception of recovery was 59 (16). The score was lower, but not significantly so, in patients with aphasia compared to patients without, both in patient (55 (15) v 65 (19), p = 0.48, Wilcoxon test) and proxy (49 (17) v 57 (18), p = 0.45, Wilcoxon test) versions of the measurement.

The combined mean (SD) physical domain recovery was 48 (16) when assessed by patients and 39 (16) when assessed by proxies. The lowest scaling success rate was for hand function and the highest for emotion domain recovery. However, during the follow up, two patients had a major depressive episode. In addition, one spouse attempted suicide (patient 8). As expected, patients with aphasia had a lower mean (SD) rate of recovery for communication (50 (37)) than those without (91 (14)), although the difference was not statistically significant (p = 0.21, Wilcoxon test).

Discussion

This study shows that the SIS measurement is applicable to patients with malignant MCA infarction 12–30 months after craniectomy. The patient’s assessment of the physical aspects of disability at 12–30 months post stroke was high (all physical domains mean recovery of 48/100). Interestingly, the proxy

Table 1: Domain scores of the SIS questionnaire filled in by seven living patients and eight proxies 12–30 months after decompressive craniectomy

| Patients/age (years)/sex | mRS version | SIS | Stren- | Hand | Mobility | ADL/ | Physical combined | Emotion | Memory | Communication | Participation | % of recovery | Stroke recovery (VAS) |
|--------------------------|-------------|-----|--------|-------|---------| IADL| Score| | | | | |
| 1/23/M | 2–95 | Patient | 60 | 0 | 98 | 85 | 61 | 100 | 95 | 100 | 61 | 75 | 75 |
| 2/49/F | 5–15 | Patient | 55 | 0 | 100 | 93 | 62 | 96 | 87 | 100 | 61 | 74 | 70 |
| 3/15/M | 2–90 | Proxy | 31 | 0 | 60 | 6 | 24 | 64 | 87 | 75 | 42 | 45 | 40 |
| 4/45/M | 4–85 | Patient | 31 | 0 | 47 | 2 | 20 | 69 | 84 | 79 | 28 | 39 | 30 |
| 5/4/F | 4–35 | Patient | 55 | 0 | 100 | 93 | 62 | 84 | 100 | 71 | 75 | 75 |
| 6/54/F | 4–35 | Proxy | 55 | 0 | 60 | 47 | 40 | 53 | 60 | 55 | 100 | 95 | 90 |
| 7/46/M | 4–60 | Patient | 55 | 0 | 68 | 53 | 40 | 89 | 70 | 40 | 27 | 47 | 40 |
| 8/46/F | 4–75 | Proxy | 55 | 0 | 28 | 33 | 20 | 44 | 37 | 29 | 24 | 25 | 10 |
| 9/50/M | 4–55 | Patient | 35 | 24 | 62 | 73 | 49 | 89 | 87 | 97 | 67 | 67 | 70 |
| 10/50/M | 4–55 | Proxy | 35 | 0 | 70 | 62 | 42 | 67 | 62 | 49 | 49 | 48 | 40 |
| Scales, mean (SD) | | | 49 (15) | (12) | 71 (21) | 61 (30) | 48 (16) | 79 (17) | 78 (17) | 67 (35) | 54 (29) | 59 (16) | 65 (18) |
| Proxies | | | 42 (12) | (0) | 62 (27) | 34 (30) | 39 (16) | 80 (16) | 72 (19) | 65 (22) | 48 (19) | 52 (17) | 47 (22) |

| Modified Rankin Scale-Barthel Index; tachyphylaxis of daily living/instrumental activities of daily living; $\Delta$combined physical score calculated from the strength, hand function, and mobility domain scores. |
| ND, not done; VAS, visual analogue scale. |
assessment of physical domains recovery was lower (39/100) than the patient assessment. In addition, the disability measured by the mRS showed that 6/8 living patients had an mRS &gt; 3, which may indicate a poor outcome. It may be that in patients with malignant MCA infarction, the patient version of the SIS overestimates the physical recovery because of cognitive dysfunction including unilateral neglect, anosognosia, or aphasia.

One main concern in malignant MCA infarction is the psychosocial impact of stroke. In our study, the percentage of recovery was good for emotion and memory but moderate for communication and partici-
pation. As expected, patients with aphasia had a lower rate of recovery for communication than patients without, though the difference did not reach statistical significance, possibly because of the small numbers. In the same way, the global percentage of recovery was lower, but not significantly, in patients with aphasia than in patients without. Interestingly, the proxy’s assessment of psychosocial recovery, though lower, was close to the patient’s assessment. In conclusion, this study shows that after aneurysmectomy for malignant MCA infarction, even though the perception of physical aspects of disability is high, that of psychosocial impairment is lower.

Open surgery for aneurysmectomy is regarded as an option when the small numbers. However, randomised trials are needed, taking into account not only death and dependency but also quality of life.

Acknowledgements
The authors thank Mapi Research Institute, Lyon, France for providing the French version of the Stroke Impact Scale 2.0 (Duncan et al, copyright University of Kansas Medical Center) and Olivier Chassany from the Département d’Anesthésie-Réanimation de la Clinique (AP-HP) for his helpful comments.

K Vahedi, J Benoist, A Kurtz, J Mateo, A Blanket, M Rossignol, P Amarenco, A Yelnik, E Vicaut, D Payen, M G Bousser

Cerebral sinus thrombosis in a patient with Cushing’s syndrome
It is well known that hypercortisolism induced by Cushing’s disease or syndrome, may be associated with other complications, including oral contraceptives, before admission. The patient had no intracranial hypertension; her fundus showed no papillo-

edema, and intracranial pressure measured by lumbar puncture was normal (4.3 mmHg). Brain computed tomogram (CT) showed a high density lesion in the left temporo-occipital lobe (fig 1A). Magnetic resonance venogram (MRV) on the first hospitalised day showed a filling defect in the left lateral sinus (fig 1B). These findings were consistent with cerebral lateral sinus thrombosis.

Laboratory data showed elevation of factor VIII (183 %, one stage clotting assay; normal range 60–160 %), VWF (275 %, normal range 60–160 %), thrombin–antithrombin III complex (15.5 ng/ml), plasminogen activator inhibitor-1 (PAI-1) (123 %), and D-dimer (2.1 µg/ml). Other major factors related to coagulation and fibrinolysis, including antithrombin III (112 %), fibrinogen (330 µg/ml), plasminogen (117 %), plasmin–

α2-plasmin inhibitor complex (0.9 µg/ml), protein C (87 %), and protein S (95 %), were within normal limits. Markers of acute phase reaction such as C reactive protein and erythrocyte sedimentation rate were not elevated. Neither antiphospholipid antibodies nor antinuclear antibodies were detected. The patient was treated with intravenous heparin and subsequent oral administration of warfarin potassium. A relative fibrinolytic enhancement following the strict anticoagu-

lation may have caused revascularisation of the lateral sinus, which was confirmed by the follow-up MRV (fig 1C). The patient’s symptoms disappeared completely.

During the extensive examination of thrombotic causes, we suspected the presence of hypercortisolism because of the presence of central obesity and moon face. As a result, we found a left adrenal tumour, which was accompanied by hypercortisolism (210 µg/l) with suppressed adrenocorticotropic hormone (3 µg/ml). The left adrenal mass showed a high uptake of 131I-iodocholesterol on scintigram. These findings were consistent with Cushing’s syndrome. After the laparoscopic left adenectomy, the patient received replacement therapy with hydrocortisone for approximately 1 year. Plasma levels of factor VIII and VWF decreased gradually to the normal level (130 % and 140 %, respectively) 1 year after adenectomy.

Discussion
We report the first case of cerebral sinus thrombosis associated with Cushing’s syn-
drome. Thromboembolic complications are well known to occur in patients with Cushing’s syndrome.1 Most are deep vein thromboses and pulmonary thromboembolisms. However, there are no reports so far to show association with cerebral sinus thrombosis and Cushing’s syndrome.

A few reports suggest that factor VIII and VWF may have roles in the development of thromboembolic complications associated with hypercortisolism.2 As well as blood group, sex, age, inflammatory, and endothe-

lial dysfunction,3 hypercortisolism is reported to be an important determinant factor for plasma levels of VWF.4 Huang et al5 showed that dexamethasone stimulated VWF release from cultured human endothelial cells. Factor VIII is mainly synthesised in the liver and secreted to the circulation. Because VWF protects factor VIII from proteases, a con-

cordant increase of factor VIII and VWF in plasma is generally observed. However, studies show that high plasma level of factor VIII (especially over 150 %) is an independent risk factor for venous thromboembolism,6 including cerebral sinus thrombosis.7 In this patient, considerable elevation of factor VIII and VWF was observed specifically before removal of the adrenal tumor. Thus, hypercortisolism may have enhanced VWF release from endothelial cells to increase factor VIII, thereby causing a hypercoagulable state. The present case also suggests that measurement of factor VIII and VWF may be useful to decide if successful adenectomy in Cushing’s syn-
drome. However, because hypercortisolism does not always cause hypercoagulable state, some genetic factors, such as polymorphism of steroid receptor, may determine whether glucocorticoids increase plasma levels of factor VIII and VWF.

It is also reported that PAI-1 is often elevated and may cause thromboembolic complications by lowering fibrinolytic activity. In our case, elevation of PAI-1 was significant but significant elevation of PAI-1 in this case may also have contributed to the thrombus formation. However, factor V Leiden, a common coagulation abnormality in Western populations, may not have partici-
pated in thrombus formation in our case, because it is considered that the mutation is not present in the Japanese population.8

Correspondence to: Dr Katayoun Vahedi, Service de Neurologie, Hôpital Lariboisière, AP-HP, 2 rue A. Paré, 75010 Paris, France; katayoun.vahedi@lrb.aphp.fr
doi: 10.1136/jnnp.2004.058537

Competing interests: none declared

References
2 Brown MM. Surgical decompression of patients with large middle cerebral artery infarcts is effective for symptom prevention. Stroke 2003; 34: 2305–6

www.jnnp.com
In conclusion, physicians should be aware that Cushing’s syndrome is a possible cause of cerebral sinus thrombosis. Plasma levels of factor VIII and VWF may play an important role in the hypercoagulable state in hypercortisolism.

S Yoshimura, T Ako, T Kitazono, T Yonekura, Y Kumai, J Kuroda, M Kamouchi, H Ooboshi, S Ibayashi, M lida
Department of Medicine and Clinical Science, Graduate School of Medical Sciences, Kyushu University, Maidashi 3-1-1, Fukuoka, Japan
Correspondence to: Dr S Yoshimura, Department of Medicine and Clinical Science, Graduate School of Medical Sciences, Kyushu University, Maidashi 3-1-1, Fukuoka, Japan; sohei-y@med.kyushu-u.ac.jp
doi: 10.1136/jnnp.2004.057315
Competing interests: none declared

References

Spinal muscular atrophy, Dandy-Walker complex, and cataracts in two siblings: a new entity?
Lower motor neurone involvement is the main feature of several neurological disorders, including the various forms of spinal muscular atrophy (SMA). A distinct form of SMA is characterised by predominantly distal weakness and atrophy of the limbs. Various combinations of SMA with neural and extraneural defects, mainly pontocerebellar hypoplasia, have also been reported.

We report a combination of distal SMA with Dandy-Walker complex and anterior polar cataracts in two brothers.

The patients were aged 25 and 23 years. Their parents, who originated from the same area of Greece, were unrelated and asymptomatic. Since the age of 10 years, both brothers presented with progressively deteriorating symmetrical distal muscle weakness and atrophy of the lower limbs, which affected mainly the anterior tibialis and peroneal muscles and, to a lesser degree, the gastrocnemius, resulting in an almost “stoat-like” appearance of the legs. Bilateral anterior polar cataracts had been diagnosed in both patients at the age of 9–11 months. Additional findings of the neurological examination in both patients were slight muscle strength reduction in both hands and forearms and decreased tendon reflexes in the upper and lower limbs, while the Achilles’ tendon reflexes could not be elicited. The plantar responses were normal. No sensory, cerebellar, or cognitive impairment was found. Dysmorphic features were not observed. The general physical examination was normal in both patients.

Extensive haematological, biochemical, and immunological investigation of both patients, including levels of creatine kinase, proactin, hexosaminidase A, anti-GM1 and anti-sialophosphatidylcholine antibodies, cortisol, thyroid hormones, vitamin B12 and folic acid, immunoglobulin G, anti-ds DNA, and antinuclear antibodies, was normal.

The electrophysiological examination revealed findings compatible with anterior horn cells involvement in both patients. Specifically, the electromyography showed chronic active denervation of distal muscles, with large amplitude motor unit potentials and presence of polyphasic potentials and spontaneous activity (fibrillations and positive waves), more pronounced in the lower limbs. Nerve conduction studies showed normal motor and sensory conduction velocities, with normal amplitudes, latencies, and F waves. No conduction blocks were recorded. Electrophysiological investigation of both parents was normal. Both patients refused consent for muscle biopsy.

Magnetic resonance imaging revealed the presence of Dandy-Walker complex in both patients. There was enlargement of the cisterna magna, with slight hypoplasia of the vermis and slight elevation of the tentorium (fig 1). No supratentorial or brainstem abnormalities were observed. The magnetic resonance imaging of the spine was normal in both brothers, as were visual and brainstem evoked responses. Ophthalmological examination confirmed the presence of anterior polar cataracts in both patients.

The karyotype was normal in both patients. Molecular genetic analysis for mutations in the survival motor neurone (SMN; exons 7 and 8 deletions), neuronal apoptosis inhibitory protein (NAIP; exon 5 and 6 deletions), and androgen receptor genes was negative.

Discussion
Our patients were two brothers with almost identical clinical and laboratory findings. One of the main features was the involvement of the anterior horn cells, which was compatible with distal SMA, according to published criteria.2 Additional features were Dandy-Walker complex and bilateral anterior polar cataracts.

There are several reports of SMA with additional features (SMA plus), among them pontocerebellar hypoplasia. These cases, however, are characterised by proximal muscles involvement, early presentation with profound floppiness at birth, mental retardation, and cerebellar signs.1 There are also rare reports of recessive distal SMA with additional features; diaphragmatic paralysis or pyramidal signs.4 None of these cases of proximal or distal SMA plus has been linked to chromosome 5q.

Familial cases of Dandy-Walker complex are not uncommon; however, the combination of the disorder with SMA seems to be quite unusual. The aforementioned severe cases of pontocerebellar hypoplasia in SMA plus clearly constitute a different nosological entity.

The coexistence of early onset cataracts with neuromuscular disorders is also unusual. Apart from the well known occurrence of cataracts in myotonic dystrophy, there are some reports of cataracts in combination with spastic paraparesis, spinocerebellar degeneration or neuropathy, and facial dysmorphism. There is also a report of familial congenital cataracts and Dandy-Walker anomaly with lissencephaly.

Figure 1 (A) Brain CT shows a high density lesion (arrow) in the left temporo-occipital lobe. (B) MRV (ECG gated two dimensional time of flight) shows a lack of flow in the left lateral sinus (arrow). (C) In the follow up MRV, the left lateral sinus (arrow) is recanalised with a residual stenosis.
We were not able to locate in the literature any reports of distal SMA in combination with any of the already described syndromes and could be the result of pleiotropy, contiguous gene syndrome, or chance. The fact that the patients were first degree relatives and presented with identical phenotypes is a strong indication that the disorder is genetically determined. With the available information on the genetics of the disorder is genetically determined. With the available information on the genetics of the main features of our patients, contiguous gene syndrome appears unlikely. We were not able to locate a genetic defect, a not altogether unexpected result, as most recessive distal SMA families remain to be genetically determined. Future investigation of similar cases should include genetic studies relevant to all three main features of the disorder.

M Panas, K Spengos, G Tsigouli, N Kalfakis, C Sfagos, D Vassiliopoulos
Department of Neurology, Eginition Hospital, University of Athens, Greece

N Markomichelakis
Department of Ophthalmology, Georgios Gennimatas General Hospital, Athens, Greece

Correspondence to: Dr M Panas, Eginition Hospital, Neurogenetics Unit, Department of Neurology, University of Athens, 74 Vas. Sofias Avenue, Athens 11528, Greece; mpanas@med.uoa.gr
doi: 10.1136/jnnp.2004.055855

Competing interests: none declared

References
Quality of life after decompressive craniectomy for malignant middle cerebral artery infarction

K Vahedi, L Benoist, A Kurtz, J Mateo, A Blanquet, M Rossignol, P Amarenco, A Yelnik, E Vicaut, D Payen and M G Bousser

J Neurol Neurosurg Psychiatry 2005 76: 1181-1182
doi: 10.1136/jnnp.2004.058537

Updated information and services can be found at:
http://jnnp.bmj.com/content/76/8/1181

These include:

References
This article cites 5 articles, 4 of which you can access for free at:
http://jnnp.bmj.com/content/76/8/1181#BIBL

Email alerting service
Receive free email alerts when new articles cite this article. Sign up in the box at the top right corner of the online article.

Notes

To request permissions go to:
http://group.bmj.com/group/rights-licensing/permissions

To order reprints go to:
http://journals.bmj.com/cgi/reprintform

To subscribe to BMJ go to:
http://group.bmj.com/subscribe/