

## Moyamoya angiopathy in Europe: the beginnings in Zurich, practical lessons learned, increasing awareness and future perspectives

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### Summary

The number of patients, especially children, diagnosed with Moyamoya angiopathy and being referred to us for treatment from all across Europe, has increased over the last few years. An increase in awareness of the occurrence of stroke in children in the general and medical population might be the main cause of this phenomenon.

Increasing awareness does not happen “spontaneously” nor does it manifest overnight!

It requires regular platforms of communication between the general population and amongst the different medical specialists mainly neurologists, paediatric neurologists, neuropsychologists, neuroradiologists, neurorehabilitation specialists, nursing staff and neurosurgeons. Presently we were lucky to conduct the first Moyamoya Symposium ever to be conducted at a European-Japanese level with participation of specialists of this particular field from across Europe and Japan.

Ever since the first child with Moyamoya was managed at the University hospital in Zurich some 7 years ago the number of patients referred to us from all across Europe increased rapidly [6–8, 11–14]. The importance of interdisciplinary communication, trust and support amongst specialists and increasing the awareness of the disease among the patients, medical personnel was and remains to be just as important as making the correct diagnosis and treatment of choice in these patients.

We present the lessons we learned during these previous years and look into the future perspectives that require our further and urgent attention.

**Keywords:** Moyamoya; angiopathy; (extracranial-intracranial bypass surgery) EC-IC bypass surgery; (superficial temporal artery to branch of middle cerebral artery) STA-MCA; (superficial temporal artery to branch of anterior cerebral artery) STA-ACA; increasing awareness.

### The beginnings in Zurich, lessons learned, increasing awareness and future perspectives

#### *The beginnings in Zurich and the lessons learned*

Sixty patients have been treated in Zurich. The majority (75%) of patients were children. The children

were sent from all across Europe and their ages ranged from 4 months to 16 years. This number of referral increased from just 1–2 patients per year in 1999 to 3–6/year in the later years. These numbers exclude the referrals and patient chart consultations performed from time to time for confirmation of diagnosis of Moyamoya in certain patients and the guideline/recommendation of a follow-up plan in certain asymptomatic patients.

Hence a systematic presurgical workup protocol was developed and used regularly to direct us in the planning of the optimal revascularisation procedure to be performed. This included a presurgical work up with clinical and neuropsychological assessment, 6-vessel cerebral angiography, transcranial Doppler, H<sub>2</sub><sup>15</sup>O-PET with a Diamox challenge. The choice of number and location of revascularisation procedures was then based mainly on the severity and extent of disease observed in the preoperative angiography and the Diamox PET studies (Fig. 1a and b).

The entire patient management could not have been possible without the close collaboration of the entire team of physicians and nurses of the children’s University hospital in Zurich, Kinderspital Zurich as well as the close collaboration of a motivated team of anaesthetists, neuroradiologists and colleagues of the Department of nuclear medicine of the university hospital in Zurich and of course all the referring physicians from across Europe.

We performed multiple direct bypass procedures of revascularisation bilaterally (STA-MCA and STA-ACA bypass) in the majority of our patients [6, 7]. Indirect revascularisation procedures (STA arteriosynangiosis, durasynangiosis) were used in combination in cases

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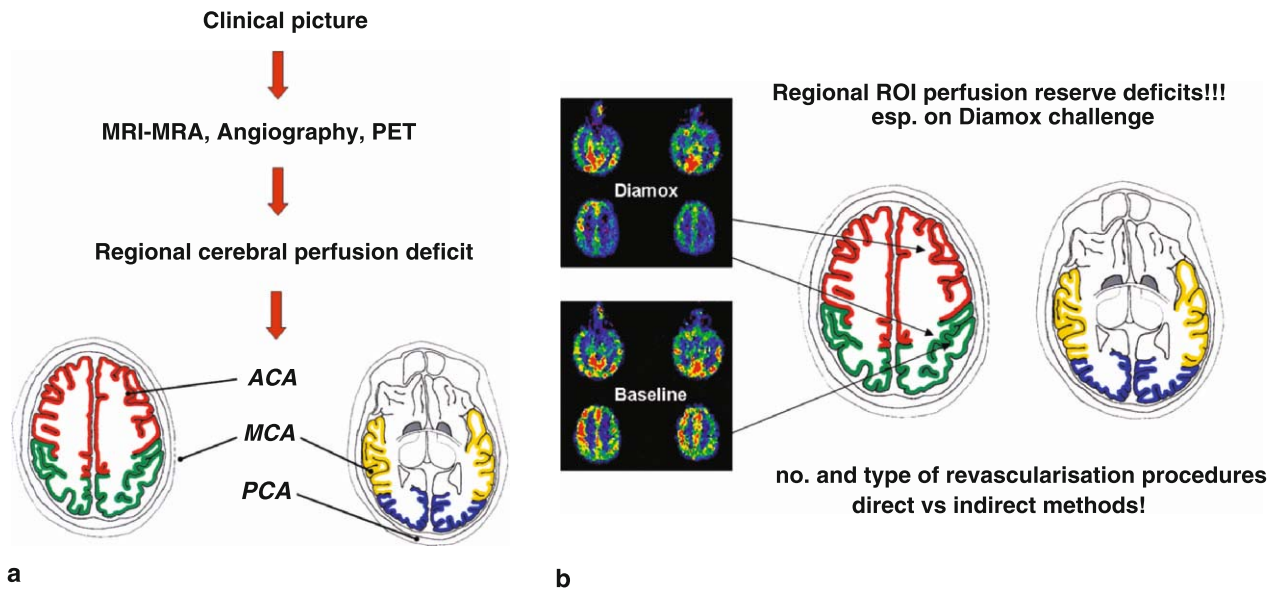


Fig. 1. (a) Scheme of preoperative evaluation: 6-vessel cerebral angiography and Diamox challenge ( $H_2^{15}O$ -PET) are the most important examinations on which the number and hence the location (arterial distribution territory of MCA middle cerebral ACA anterior cerebral or PCA posterior cerebral artery) of revascularisation procedures (STA-MCA, STA-ACA or PCA bypass) depends upon. (b) Regional ROI (region of interest) perfusion reserve deficits are demonstrated

where the donor or recipient arteries were not available or were of inadequate calibre for the anastomosis. The postoperative follow-up showed no further stroke and improvement in cognitive functions in children who demonstrated a preoperative frontal lobe executive functional impairment.

Reviewing the American and Japanese literature [1, 5, 9] although STA-MCA bypass procedures are performed frequently for Moyamoya angiopathy, little importance is given to frontal brain reperfusion, therefore only few STA-ACA bypass are being performed [3, 4, 10], especially in children.

The need to reperfuse the frontal brain regions is extremely important in preventing mental retardation in children before the age of 5 years. Hence frontal brain hypoperfusion especially in the dominant side guided us to perform STA-ACA bypass procedures. Also the importance of pre- and postoperative neuropsychological assessment especially of the frontal brain regions in children should be incorporated in the surgical management.

#### *Increasing awareness and future perspectives*

##### Children's book project

A project of increasing awareness of Stroke prevention and Moyamoya angiopathy was undertaken end of 2005

and completed in 2006. Apart from regular lectures and presentation on the topic of Moyamoya and especially stroke in children regionally and internationally, a children's book project was initiated and completed in August 2006. Two books "Sven" and "Fatma's fantastic journey" were launched to explain the disease to the parents, children, affected patients as well as the medical personnel managing them. These books have been published in English and German and further sponsoring is required and underway to publish them in French, Italian and Dutch. Sven will be published in Japanese soon (Fig. 2).

##### Self-help group on Moyamoya

The first Moyamoya self-help group was also initiated in May of 2006 and parents, children and patients met with us to outline the problems faced during the period of securing a diagnosis of Moyamoya where up to 3 years of waiting had been lost, getting the diagnosis, finding a specialist and a hospital providing the infrastructure for managing and treating this illness.

##### Epidemiological study

The last epidemiological study on the number of patients diagnosed in Europe was performed in 1996 [2, 13]. It is time to perform a European epidemiologi-



Fig. 2. Children's book project: written and published by the first author in relation to a "Moyamoya awareness" Project carried out by the first author. Sven: a children's book for children up to 7 years of age defining the Moyamoya angiopathy, the diagnosis and preoperative investigation methodology and the bypass procedure. This book helps in eliminating the anxiety the children and parents suffer from when the term "Moyamoya" first falls on the scene. Fatma's fantastic journey, a comic for children above 7 years of age: Sven returns to Zurich for his follow-up examinations and meets Fatma and two other children suffering from Moyamoya. Together they make an adventurous journey into the human brain

cal study to find out the burden of this disease here in Europe and to outline the different management protocols (conservative vs neurosurgical) being followed. A consensus to perform such a Questionnaire was reached during the Moyamoya Symposium and will soon be underway.

### Concluding remarks

Creating and managing a practical working environment for diagnosis, preoperative evaluation, surgical treatment, regular postoperative follow-up, patient and family support along with continuous awareness, education and research is what is required in the future in Europe to provide for the optimal care for the European Moyamoya patients especially children. Also the importance of basic and specialised continuous microsurgical training in the lab has to be a must in the neurosurgical training programmes across Europe motivating young neurosurgeons in the field of vascular neurosurgery.

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