Diagnosis of Moyamoya Angiopathy
Dear parents,

Finding out that your child has been diagnosed with Moyamoya Angiopathy can create a difficult situation for the entire family. It is precisely at such a moment that a ray of hope can be given by telling you that this neurological disorder is treatable and Moyamoya children can resume their normal lives.

The Moyamoya Center at the Children’s Hospital Zurich is one of the few competence centres in the world that specialises in Moyamoya Angiopathy. Children receive age-appropriate medical care in line with the young patients’ needs combined with close personal support. We place a high priority on ensuring that children and their families feel well taken care of, both today and in the future. For this reason, support is usually not terminated when the convalescence period comes to an end but rather continues up until the children become adults.

Our mission is to put smiles back on the faces of our young Moyamoya patients.

Nadia Khan, MD, Director Moyamoya Center

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What is Moyamoya Angiopathy?

This neurological disorder is a little-known disease in which the main brain arteries supplying the brain with blood supply narrow and close. The brain tries to compensate for impaired circulation by forming cloud-like structures (collateral arteries) called the 'Moyamoya' arteries. Japanese neurosurgeons J. Takeuchi and K. Shimizu, who first described the symptoms in 1957, named this disease 'Moyamoya' or 'puff of smoke' due to the resemblance of these collaterals to clouds of smoke.

Moyamoya can and must be treated, otherwise, the narrowing brain vessels can lead to decreased blood supply to the brain resulting in loss of motor function, speech disorders, repeated strokes resulting in severe physical and mental handicap. Blood circulation is restored through bypass operations. This prevents further attacks and your child can then lead a normal life once more, without mental or physical impairment.
Diagnosis and preliminary examinations
Moyamoya Angiopathy is diagnosed in your child using imaging techniques such as an MRI scan (Magnetic Resonance Imaging). Revascularisation operations are the only treatment option for restoring normal blood circulation in Moyamoya patients. Therefore, a hospital stay for your child should be initiated.

Hospital admission
During the first day at the Children’s Hospital Zurich, an admission meeting with the ward doctor is first on the schedule, followed by clinical, neurological, developmental paediat-
rics and genetic clarifications. Nadia Khan will then conduct an in-depth discussion with you, your child and other family members. Do not hesitate to ask any questions that remain unanswered.

Anaesthesia is required for the subsequent neuroradiological exams, including an examination of the brain structure and the six main blood vessels of the brain using MRI and angiography as well as an evaluation of the blood flow with a PET scan (Positron Emission Tomography). Why does your child have to be anaesthetised? For one thing, the young patients must lie still for a long period. For another, the strange and highly specialised equipment could cause unnecessary stress for children. The examinations are carried out quickly and practically unnoticed thanks to anaesthesia. In contrast to an older person, a child’s body can quickly and completely recover from the anaesthetic, even when faced with repeated sedations. Additional specific examinations, which will provide important medical information for the upcoming surgical intervention, may also be needed depending on the extent of the angiopathy.

Planning the revascularisation operations

Based on the results of all the preliminary examinations, meticulous planning for the surgical intervention can begin. Another parent meeting will be scheduled at this point, which is also attended by the anaesthesiologist. Everyone will have the opportunity to discuss the operation in detail and questions can be answered. You may also visit the intensive care ward with your child. Becoming acquainted with this ward helps young patients to overcome their fears and satisfy their curiosity. Before the operation, your child needs to continue taking the prescribed aspirin medication and it is important for your child to drink at least two litres of fluids daily. On the day before the operation, it is advised to wash your child’s hair for purpose of hygiene.

The day of the operation

On the operation day, you may accompany your child on the trip from their hospital room to the anaesthesia delivery room. As soon as the anaesthetic begins to take effect and your child is asleep in the competent hands of our anaesthesia team we
will ask you to leave and be patient as the operation will last several hours. During this period we will keep you informed about the progress of surgery, so the best is to leave the hospital area and visit the city and keep yourself busy. As soon as the operation is completed, your child will be transferred to the intensive care ward for the reawakening process and monitoring, and you may then visit at a time suggested to you by the neurosurgeon. After spending a night at the intensive care unit after stabilisation your child may return to their room. Prior to transfer back to the room a routine CT scan will be performed. This will rule out the possibility of any complications such as bleeding or strokes. With a clear unremarkable CT the day after the operation, your child will once again be able to get up and play. In the days following the operation, your child’s face and head may become quite swollen. This swelling is customary and disappears after two to four days. However, it is vital to ensure that the head section of your child’s bed is kept at a 30- to 45-degree angle during the entire hospital stay to reduce the swelling.

Post-operative examinations and discharge from the hospital
Depending on the number of operations required, the duration of the hospital stay varies between seven and fourteen days after surgery. During this time, your child has the opportunity to recover from the surgical intervention. On the fourth day after the operation, the surgical wound will be examined and redressed. The stitches do not need to be removed since they will dissolve completely during the following eight weeks. Regular follow-up examinations during the hospital stay will confirm that the recovery is progressing.

Ambulatory follow-up
If no complications arise, the first follow-up examination will take place around six months after the final operation. Clinical and neuroradiological examinations such as an MRI, PET scan and angiography will be scheduled. Depending on the clinical follow-up, additional examinations will be carried out annually or every three years until puberty.
Thorough preparation makes your child’s hospital stay easier

The upcoming hospital stay often causes a child to experience insecurity and fears. You can reassure your child with age-appropriate clarifications. Explain to your child why he or she needs to visit the Children’s Hospital Zurich and what happens there. In addition to this brochure, the Moyamoya Center has published two books to assist you with such conversations:
- “Sven”, a picture book for children 4 and older
- “Fatma’s Fantastic Journey”, a comic book for children 10 and older

Both books can be requested for free from the Moyamoya Center at the Children’s Hospital Zurich.

A visit to the hospital beforehand can also reduce worries and may even create healthy curiosity and help to answer the questions of the entire family. For this reason, the Children’s Hospital organises regular hospital tours. The automatic telephone recording at +41 44 266 80 88 will inform you when the next tour is to take place.

You can help to advance the treatment of Moyamoya

The Moyamoya Center is committed to further researching this neurological disorder and continuing the constant development of treatment methods. Every single Moyamoya child profits from the latest findings and methods.

However, we depend on the cooperation of our patients to carry out this research. Have you ever considered allowing your child to participate?

Nadia Khan, MD, and her nurse coordinator would be happy to explain the current research projects and all other aspects related to participation during a personal meeting with you.
Welcome to the Moyamoya Group

Patients at the Moyamoya Center and their relatives are organised into a contact group. The main objective of the network is to exchange information with like-minded people – either in a group or on a one-to-one basis. All interested individuals meet together at least once a year for an information and sharing event.

You can get to know members from all over the world. The Moyamoya Center is happy to provide you with contact details.

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Additional information about Moyamoya Angiopathy
- Website: www.moyamoya.net
- Specialist textbook: "Moyamoya Disease, Diagnosis and Treatment" by John E. Wanebo, Nadia Khan, Joseph M. Zabramski and Robert F. Spetzler, published by the Thieme-Verlag. ISBN no. 978-1604067309