Multiple bur hole surgery for the treatment of moyamoya disease in children

CHRISTIAN SAinte-Rose, M.D., RICARDO OLIVEIRA, M.D., STÉPHANIE PUget, M.D., LIANA BENI-ADANI, M.D., NATHALIE BODDAERT, M.D., JOHN THORNE, F.R.C.S., ALISON WRAY, F.R.A.C.S., MICHEL ZERAH, M.D., AND MARIE BOURGEois, M.D.

Department of Pediatric Neurosurgery, Hôpital Necker–Enfants Malades, Paris, France; and Department of Neurosurgery, Royal Children’s Hospital, Melbourne, Australia

Object. The authors’ aim in this study was to review their experience in the use of indirect revascularization alone in a series of 14 children with moyamoya disease, in which numerous bur holes and arachnoid openings were made over each affected hemisphere.

Methods. Revascularization through multiple bur holes and arachnoid openings was performed in 14 children (mean age at diagnosis 6.5 years [range 3–15 years]) who suffered from progressive moyamoya disease. The authors performed surgery in a total of 24 hemispheres during 18 procedures. Ten children underwent bilateral multiple bur hole procedures, three underwent a unilateral procedure in the more severely affected hemisphere, and one child had previously undergone an encephaloduroarteriomyosynangiosis on the contralateral side. Ten to 24 bur holes were made in the frontotemporoparietooccipital area of each hemisphere, depending on the site and extent of the disease. Early postoperative perfusion magnetic resonance imaging studies, performed in the five most recent cases, showed restoration of cortical perfusion as early as 3 months, which was confirmed on subsequent angiography studies (performed between 8 and 12 months postoperatively) that showed excellent revascularization of the ischemic brain by external carotid artery collateral vessels. None of the children sustained further ischemic attacks postoperatively. Motor improvement was noted in those who had presented with paresis. A single seizure episode occurred in two patients at 2 weeks and 5 months after surgery; both children had presented with epilepsy. There were no postoperative deaths, and only one complication (an infected lumbar shunt in the patient who required cerebrospinal fluid [CSF] drainage). Five of the 18 procedures were complicated by subcutaneous CSF collections, which resolved with tapping and compressive head dressings; a transient lumbar drain was necessary in one case.

Conclusions. The results obtained in this series suggest that in children with moyamoya disease this simple technique is both effective and safe. Furthermore, it is effective as a sole treatment without supplementary revascularization procedures.

KEY WORDS • moyamoya disease • indirect revascularization • bur hole • pediatric neurosurgery

MOYAMOYA disease is a chronic progressive cerebrovascular disease in which an abnormal vascular network and collateral blood supply develop following stenosis of the distal intracranial ICAS.1,7,11,16,18 In children, moyamoya disease is usually accompanied by TIAs or a complete stroke, although the presence of seizures is not uncommon. In contrast to adults, children rarely suffer hemorrhage. Several surgical techniques have been used in which revascularization of the ischemic regions of the brain is attempted. Both direct revascularization through STA–MCA anastomosis and indirect methods such as EMS, omental flap, EDAS, EDAMS, and pial synangiosis have been performed with varying success.1,8,10,12,13 Combinations of direct and indirect revascularization techniques have also been attempted.2,5,15

In 1989 Endo, et al.,1 observed marked neovascularization occurring across a frontal bur hole, which had been drilled to allow external drainage of an intraventricular hemorrhage in a child with moyamoya disease. The same group later used frontal bur holes in combination with EMS in the treatment of moyamoya disease in children and showed that this led to better revascularization of the frontal area compared with EMS alone. A few years later, Kawaguchi and colleagues4 performed the bur hole procedure without addi-
Patient Population

We reviewed a consecutive series of children who presented to Hôpital Necker-Enfants Malades with ischemic symptoms and imaging-confirmed moyamoya disease. Fourteen children, ranging in age from 3.5 to 16 years at the time of surgery, with symptomatic progressive moyamoya disease underwent the revascularization procedure. Preoperatively all children underwent a neurological examination, electroencephalography, CT scanning, MR imaging, and angiography; in the most recent cases a preoperative perfusion MR image was also obtained. In cases of staged surgery, we initially treated the more severely affected hemisphere. In 13 patients, the multiple bur hole procedure was the only revascularization operation undertaken. In one child, however, a previous EDAMS procedure had been performed in the right hemisphere, and the subsequent bur hole procedure was performed 18 months later in the left hemisphere. In all 24 operations, multiple bur hole placement was the sole revascularization procedure. All children underwent clinical follow-up examinations after surgery. Magnetic resonance imaging was performed at 3, 6, and 12 months postoperatively, and yearly thereafter. Postoperative angiograms were obtained at 6 months and 2 years postsurgery. In later cases, perfusion MR images were obtained.

Surgical Technique

After the induction of general anesthesia, the patient is placed supine. In cases in which a unilateral approach alone is used, the head is turned to the contralateral side. For bilateral revascularization procedures, the head is kept neutral and flexed (as in scaphocephaly surgery) to achieve good exposure of the entire calvaria. As is routine in our unit, a single dose of an antibiotic agent is administered at the beginning of the procedure. A 1-cm strip of hair is shaved in preparation for the skin incision. The skin is infiltrated in the subgaleal plane with saline solution for ease of dissection. The skin incision for this procedure evolved over the course of this study. We used a retrocoronal opening crossing the midline, initially a linear incision, and later a zigzag incision as is our preference for cosmesis. The retrocoronal incision allows bilateral exposure of the frontal, parietal, temporal, and partial occipital regions. For unilateral procedures a T-shaped incision (a unilateral coronal incision plus a midline incision) is used to preserve the contralateral vascularization.

The galea is carefully elevated, paying meticulous attention to the dissection and preserving scalp vascularization as much as possible. The periosteum is left in place on the bone, preserving the vessels that will form the future collateral network. Multiple triangular incisions are made in the periosteum and elevated as small flaps to expose the bone. The openings are placed 3 cm apart over the entire exposed calvaria, covering the involved vascular territories (Fig. 1). The medial limit is 3 cm from the midline to avoid bleeding resulting from injury to the sinus or the draining veins. Bur holes are then made at each exposed area with a high-speed drill fitted with a ball-shaped tool. Using the surgical microscope the dura mater is opened through each bur hole while branches of the meningeal arteries are avoided (Fig. 2). The arachnoid mater and pia mater are then nicked just enough to ensure a true opening but not to cause significant bleeding. Hemostasis is achieved with cottonoid patties and gentle saline irrigation. Cautery is avoided as much as possible to preserve the potential anastomotic vessels. The previously elevated periosteal flap is then placed in contact with the exposed brain through each bur hole. The galea is carefully repositioned, and a two-layer watertight closure of the skin is performed. A compressive head dressing is applied for 4 to 5 days.

On the 1st postoperative day plain skull x-ray films (anteroposterior and lateral) are obtained to show the location of the bur holes; these images can be compared with findings on subsequent angiograms (Fig. 3). A CT scan of the brain is also obtained to ensure that there is no unexpected bleeding or CSF collection.

At the beginning of our experience the scalp flap was elevated in the subperiosteal plane. However, even though it did not affect the revascularization procedure, we abandoned this step because the blood loss was greater and the subcutaneous CSF collection more problematic.

Results

Eighteen revascularization procedures (four unilateral,
Multiple bur holes in moyamoya disease

Fig. 2. Schematic drawing of a bur hole.

Fig. 3. Preoperative (left) and postoperative (right) skull x-ray films obtained in a child treated with multiple bur holes.

eight staged bilateral, and six single-setting bilateral) via multiple bur holes and arachnoid openings were performed in 14 children with moyamoya disease between April 1999 and June 2004 at the Hôpital Necker–Enfants Malades, Paris. The relevant clinical characteristics of this series are reported in Table 1. Ten children underwent bilateral surgery in which multiple bur holes were placed either in a staged fashion (four cases) or during the same procedure (the six most recent cases), and three children underwent a unilateral revascularization procedure. One child had previously undergone a right-sided EDAMS procedure 18 months before the multiple bur hole operation was performed on the left side. The delay between the two operations in children undergoing staged bihemispheric surgery ranged from 3 to 5 months.

The children presented with both ischemic symptoms (TIA and stroke) and nonspecific symptoms (headache and seizures). According to the clinical grading of moyamoya disease by Matsushima and colleagues, the status in seven children was Stage IV (initial infarction followed by a TIA), that in six was Stage III (recurrence of a TIA with imaging evidence of infarction or neurological deficit), and that in one was Stage II (a TIA with no neurological deficit or evidence of infarction on imaging). All preoperative CT and MR imaging studies revealed abnormal findings, demonstrating the features of moyamoya disease with intracranial carotid artery stenosis, impaired distal circulation, and varying degrees of compensatory basal anastomotic vessels in association with ischemic areas and infarction or brain atrophy. The angiographic findings, shown in Table 1, were graded according to the classification of Suzuki, et al. (Stage 2, carotid artery bifurcation stenosis with initial basal moyamoya vessel formation; Stage 3, arterial narrowing of the circle of Willis and progression of basal moyamoya vessels; and Stage 4, progressive narrowing of the main cerebral arteries with almost complete disappearance of the posterior cerebellar artery, minimal residual patency of basal vessels, and the development of extracranial collateral vessels).

In our study there were no intraoperative deaths. The duration of the surgical procedure ranged from 70 to 150 minutes, depending on the number of bur holes placed (range 10–24). Blood loss was minimal, and no child required an intraoperative blood transfusion.

Subcutaneous CSF effusions were commonly observed on the postoperative CT scans but were minimal and usually successfully treated by compressive head dressings. In four patients aspiration was also necessary. In one case the collection persisted, and a lumbar shunt was inserted. Unfortunately this procedure was complicated by a shunt infection that necessitated removal of the device; the prior period of drainage, however, was sufficient to resolve the subcutaneous collection.

To date we have between 25 months and 5.6 years (mean 4.2 years) worth of clinical follow-up data for all patients. All 14 children exhibited distinct improvement in their daily functioning and school performance. Of the five children who experienced seizures preoperatively, a single seizure occurred in two at 2 weeks and 5 months postoperatively. For all nine children with preoperative paresis the motor deficit improved significantly but did not fully resolve. No new ischemic event (TIA or stroke) occurred in any patient.

All 14 postoperative angiograms demonstrated excellent revascularization by ECA collateral vessels at the sites of the bur holes, whereas the stenotic changes of the primary disease continued to progress (Figs. 4–6). The angiographic outcome remains difficult to quantify in the absence of a validated scoring systems. Matsushima, et al. described a post-EDAS score based on four features: dilation of the arterial territories (number of visible arteries: −1, decrease; 0, no change; 1, slight increase; 2, marked increase; and increase in the diameter of the three main arteries: 0, no change; 1, < 30%; 2, > 30% to a maximum score of 3); dilation of the donor artery (0, no change; 1, < 30%; 2, 30–50%; and 3, > 50%); area of brain revascularization (0, no visualization; 1, < 30 mm²; 2, 30–60 mm²); and 3, > 60 mm²; and decrease in the moyamoya vessels (anterior and posterior circulations scored: −1, increase; 0, no change; 1, slight change; and 2, marked decrease to a maximum score of 3). Given the lack of other postoperative scoring systems we used the Matsushima staging system in this series (Table 1). In another article the same senior author proposed a grading system for postoperative arteriographic results based on the classification of the development of collateral formation demonstrated on the postoperative external carotid angiograms: A, more than two thirds of the MCA distribution; B, between two thirds and one third; and C, slight or none. If one were to use this system, one would find that seven formations were classified as A, 14 as B, and three as C. However, the value of this classification to assess the effec-
tiveness of a given revascularization technique is debatable as it is evident that the collateralization (after indirect technique) occurred initially in the most severely affected areas. In other words, this classification is influenced both by the extent of the preoperative ischemic brain areas and the effectiveness of the revascularization technique.

Despite progression of the stenotic changes, all hemispheres in which multiple bur holes were placed showed ex-
cellent revascularization. Interestingly in the child in Case 1 a marked difference was observed between the score for the left hemisphere that had previously been treated with an EDAS and the score for the right hemisphere treated with subsequent multiple bur hole placement.

The functional effectiveness of the neovascularization was confirmed by perfusion MR imaging in the more recent cases (Fig. 7). Perfusion MR imaging demonstrated an increase in the cerebral blood flow of the previously ischemic brain areas as early as 3 months postoperatively, and before the collateral network from the ECA become visible on angiography.

Discussion

In this paper, we describe a method of making multiple bur holes and arachnoid openings in 24 hemispheres for treating children with moyamoya disease. All postoperative angiograms demonstrated excellent results of cortical revascularization at sites corresponding to the bur holes. Clinically all children exhibited some improvement in their motor function (if dysfunction was present preoperatively), seizures, or TIAs, and no child suffered an ischemic event postoperatively. The principle of indirect revascularization in patients with moyamoya disease is based on the natural
ability of these collateral vessels to develop in these patients and on the fact that revascularization of the ischemic brain areas can be achieved from an extracranial (ECA) blood supply. This process is enhanced by establishing a surgical connection between the surface of the brain and tissue, which is supplied by the ECA and which is rich in vasculature (galea, temporal muscle, and dura). Previous authors have demonstrated that the driving force for the collaterization is the ischemic cortex. Studies of CSF samples obtained in patients with moyamoya disease have shown changes in the CSF cytokines that act in an angiogenic fashion. This correlates well with the finding in our series that the most ischemic areas were the first to develop anastomotic circulations. Reviewing the elegant description by Suzuki, et al., of the natural history of moyamoya disease observed on angiography, we note that the final stage is attempted revascularization of the cortex with multiple ECA anastomoses. The placement of multiple bur holes has an empirical basis in revascularization as it allows the brain to form these collateral vessels earlier, thereby protecting the child from the traditionally large number of ischemic events.

The principle that effective revascularization can be achieved using a bur hole technique, without additional procedures, has already been demonstrated by Kawaguchi, et al. In a series of 10 adult patients in whom between one and four bur holes were placed over each hemisphere, good revascularization was achieved in 41 of 43 areas underlying the bur holes. In our series, we used the same method in a pediatric population, but greatly increased the number of bur holes (10–24 bur holes in each hemisphere) so that extensive coverage of the hemisphere could be achieved. In all patients bur holes were drilled in the frontoparietotemporal areas, occasionally extending to the occipital area depending on the preoperative angiographic filling deficits. We believe this matching of revascularization to all the ischemic territories is a fundamental benefit of this technique and that the reason for the failures of other techniques reported in the literature is related to their limited areas of revascularization. In STA–MCA anastomosis, as well as in EMS, EDAS, and pial synangiosis, good neovascularization may be seen in the temporal and parietal regions; however, in many patients there are angiographic and clinical signs of other ischemic territories, especially with disease progression. Accordingly, both STA–MCA and EMS operations have been shown to improve mainly symptoms related to the MCA territory (aphasia and motor and sensory deficits). Leg weakness, abnormal mentation, and visual field abnormalities due to ischemia in areas supplied mainly by the anterior and posterior cerebral arteries did not improve.

Although for direct revascularization (STA–MCA anastomosis) it is true that immediate collateral flow may be attained, its benefit is questionable because the clinical outcome is no different from that achieved with an indirect revascularization procedure. One has to remember also that this operation may be technically difficult to undertake in young children. Temporary occlusion of the recipient MCA branches may disrupt important leptomeningeal collateral networks, and proximal occlusive changes may limit collateral flow, which is intended to be diverted to the ischemic brain. It is therefore encouraging that in this study we have demonstrated an early improvement in cerebral perfusion prior to the previously described imaging improvement. It is important to emphasize that the children in this study were not selected for the indirect revascularization procedure based on the severity of the clinical presentation at admission because it is our practice to place multiple bur holes in all patients with moyamoya disease.

In children with moyamoya disease, EDAS, other indirect revascularization methods, or combined procedures may be effective, but the extent of the revascularization has been shown to be dependent on the area and size of contact between the extracranial tissue and the brain. This led Houkin, et al., to suggest that as much brain as possible be exposed, and the external tissues be applied as widely as possible. Matsushima, et al., recently reported an increased incidence of revascularization after using a combination of EDAS, EMS, and EMS in 12 patients (16 hemispheres), compared with EDAS alone. These authors reported good revascularization in 72% of the cases based on postoperative angiograms, and TIA's disappeared in 44% of children in 90% of children at 6 months postsurgery. These reports suggest that extensive surgery is required to yield better results. The clear disadvantages of such procedures performed in children are the large craniotomy, the possible blood loss from an operation of such magnitude, and the fact that the frontal or occipital areas do not revascularize. Postoperative strokes and TIA's have been reported following these procedures, which probably represent ongoing ischemia. It has been suggested that a combined technique of direct and indirect
Multiple bur holes in moyamoya disease

surgery may be superior, resulting in better and more extensive revascularization. The method which we have described has the following advantages. 1) Multiple bur holes may be placed over the frontal, parietal, temporal, and occipital areas as necessary, whereas in other approaches it is difficult to genuinely revascularize the entire hemisphere even with an extensive craniotomy. 2) The procedure is short and safe, and there is minimal blood loss; thus it may be an excellent treatment option even in high-risk cases. 3) In this study, we have placed supine (head elevated 30°). In a recent case (not included in this series) multiple small linear incisions (four on each side) were made in a severely debilitated child in an attempt to further minimize blood loss, preserve the preexisting collateral network as much as possible, and reduce the risk of postoperative subcutaneous fluid collection.

Conclusions

The technique which we have described has the following advantages. 1) Multiple bur holes may be placed over the frontal, parietal, temporal, and occipital areas as necessary, whereas in other approaches it is difficult to genuinely revascularize the entire hemisphere even with an extensive craniotomy. 2) The procedure is short and safe, and there is minimal blood loss; thus it may be an excellent treatment option even in high-risk cases. 3) In this study, we have shown that in the pediatric population, surgery involving the drilling of multiple bur holes results in good collateral revascularization, improved cerebral perfusion, and a dramatic reversal of clinical ischemia. 4) Finally, this technique does not preclude further surgical interventions if a patient should require them as an adult.

References


Manuscript received May 12, 2005. Accepted in final form September 5, 2006.

Address reprint requests to: Christian Sainte-Rose, M.D., Service de Neurochirurgie Pédiatrique, Hôpital Necker–Enfants Malades, 49, rue de Sèvres, Paris 75743, Cedex 15, France. email: christian.sainte-rose@rck.ap-hop-paris.fr.