Indirect revascularization for Pediatric Moyamoya disease

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In children, MMD frequently manifests as ischemic symptomatology. Cerebral perfusion gradually decreases as the disease progresses, which often leads to cerebral infarction. The benefits of revascularization surgery, whether direct or indirect, have been well-established in MMD patients with ischemic symptoms. In adults, the increase in cerebral blood flow achieved with indirect revascularization is often unsatisfactory, and direct revascularization is usually feasible. In children, however, direct revascularization is frequently technically not feasible, whereas the response to indirect revascularization is excellent, although 1 or 2 weeks are required for the stabilization of symptoms. The authors describe surgical procedures and perioperative care in indirect revascularization for MMD.¹⁾.

Indirect revascularization is one of the surgical treatment options used for pediatric Moyamoya disease. It involves creating new blood vessels to bypass the narrowed or blocked arteries in the brain, promoting better blood flow and oxygen supply to the affected areas. This procedure is particularly useful when direct revascularization is not feasible due to the lack of suitable donor vessels or other medical considerations.

There are two main techniques for indirect revascularization in pediatric Moyamoya disease:

Encephaloduroarteriosynangiosis (EDAS).

see Encephaloduroarteriosynangiosis for Pediatric Moyamoya disease.

The indirect revascularization procedures are typically considered when the blood vessels are too small or fragile for direct bypass surgery or when there is limited availability of suitable donor vessels. Indirect procedures are also used for bilateral cases of Moyamoya disease (both sides of the brain are affected), as the direct method might not provide sufficient blood flow for both sides simultaneously.

It is important to note that while indirect revascularization can help improve blood flow, the formation of new blood vessels is a gradual process that takes time. The newly formed vessels may not fully mature until several months after the surgery, so close monitoring and follow-up are crucial to assess the effectiveness of the procedure and manage any potential complications.

Complications

As with any surgical procedure, there are risks associated with indirect revascularization, and the decision to pursue this approach should be made after a thorough evaluation by a skilled pediatric neurosurgeon or cerebrovascular specialist. The ultimate goal of the surgery is to reduce the risk of stroke and improve the patient's neurological function and quality of life.

Surgical revascularization decreases the long-term stroke risk in children with moyamoya disease but can be associated with an increased risk of stroke during the perioperative period. Evidence-based approaches to optimize perioperative management are limited and practice varies widely. Using a modified Delphi process, Sun et al. sought to establish expert consensus on key components of the perioperative care of children with moyamoya undergoing indirect revascularization surgery and identify areas of equipoise to define future research priorities.

Thirty neurologists, neurosurgeons, and intensivists practicing in North America with expertise in the management of pediatric moyamoya were invited to participate in a three-round, modified Delphi process consisting of a 138-item practice patterns survey, anonymous electronic evaluation of 88 consensus statements on a 5-point Likert scale, and a virtual group meeting during which ideas were discussed, revised, and reassessed. Consensus was defined as \geq 80% agreement or disagreement.

Thirty-nine statements regarding perioperative pediatric moyamoya care for indirect revascularization surgery reached a consensus. Salient areas of agreement included the following:

(1) children at high risk for stroke and those with sickle cell disease should be preadmitted prior to indirect revascularization;

(2) intravenous isotonic fluids should be administered in all patients for at least 4 h before and 24 h after surgery;

(3) aspirin should not be discontinued in the immediate preoperative and postoperative periods;

(4) arterial lines for blood pressure monitoring should be continued for at least 24 h after surgery and until active interventions to achieve blood pressure goals are not needed;

(5) postoperative care should include hourly vital signs for at least 24 h, hourly neurologic assessments for at least 12 h, adequate pain control, maintaining normoxia and normothermia, and avoiding hypotension, and (6) intravenous fluid bolus administration should be considered the first-line intervention for new focal neurologic deficits following indirect revascularization surgery.

In the absence of data supporting specific care practices before and after indirect revascularization surgery in children with moyamoya, this Delphi process defined areas of consensus among neurosurgeons, neurologists, and intensivists with moyamoya expertise. Research priorities identified

include determining the role of continuous electroencephalography in postoperative moyamoya care, optimal perioperative blood pressure and hemoglobin targets, and the part of supplemental oxygen for treatment of suspected postoperative ischemia².

Headaches often persist or newly develop after revascularization surgery in MMD patients. Accompanying nausea or vomiting and occurrence upon awakening are characteristic features. Postoperative headache does not necessarily imply insufficient disease control ³⁾

Case series

A retrospective review of patients with MMD who underwent bifrontal indirect bypass surgery was performed. Clinical features, perioperative data, and angiographic, perfusion, and functional outcomes were compared between the 2 groups. Propensity score matching was performed to compare the perioperative characteristics and clinical outcomes.

Results: A total of 346 patients were included in this study, 111 patients underwent bifrontal craniotomy EGPS, and 235 patients had bifrontal multiple burr hole EGPS. An insignificant higher rate of postoperative infarction (11.7% vs 5.5%, P = .072) and more postoperative hemorrhage occurred in the craniotomy EGPS group (3.6% vs 0%, P = .004). Of the 83 patients selected with propensity score matching for each group, the duration of operation was shorter (P < .001) and the amount of intraoperative bleeding was significantly less in the multiple burr hole EGPS group (P = .008). There was no difference in clinical outcomes between the 2 groups.

Conclusion: Bifrontal multiple burr hole EGPS has benefits over craniotomy with shorter surgical time, less intraoperative bleeding, fewer postoperative complications, and comparable perfusion and functional outcomes. Multiple burr hole EGPS is a safe and effective method that might be considered for revascularization of the anterior cerebral artery territory in pediatric patients with MMD⁴⁾.

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