Choroid plexus coagulation in infants with extreme hydrocephalus or hydranencephaly

Clinical article

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Object. Severe hydrocephalus and hydranencephaly are common congenital conditions in Kenya. In patients with these conditions, ventriculoperitoneal (VP) shunts are associated with appreciable complications and endoscopic third ventriculostomies (ETVs) have limited success. Endoscopic choroid plexus coagulation (CPC) to diminish CSF production is a potential treatment option. The purpose of this study was to evaluate the effect of CPC without ETV in infants with severe hydrocephalus or hydranencephaly.

Methods. Medical records of infants with severe congenital hydrocephalus or hydranencephaly who underwent CPC in Kijabe Hospital from November 2010 to April 2013 were reviewed retrospectively. Thirty-three patients with complete medical records and preoperative radiographic images were identified. After CPC, the infants were followed in the Kijabe Hospital outpatient department, in mobile clinics, or by telephone. Success of the CPC was defined as resolution of preoperative symptoms, stabilization of head size, and avoidance of VP shunt placement.

Results. Patients were followed from 30 to 608 days (median of 120 days). Three patients were lost to follow-up. Of the 30 evaluable patients, CPC was considered to be successful in 13 (43.3%), including 8 of 20 patients with severe hydrocephalus and 5 of 10 with hydranencephaly. Failure of CPC was evident from increased head circumference in 14 (82%) of 17 patients and from CSF leakage in 3. Of the 17 failures, 13 occurred within 3 months of surgery. Six patients died: 3 whose CPC procedures were failures, 2 whose CPC was successful, and 1 postoperatively. Of the 17 in whom CPC failed, 10 subsequently underwent VP shunt insertion.

Conclusions. CPC stabilizes macrocephaly in approximately 40% of infants with severe congenital hydrocephalus and hydranencephaly and can be considered as an alternative to VP shunt placement.

Key Words • hydrocephalus • hydranencephaly • choroid plexus coagulation

hydrocephalus is the most common neurosurgical condition presenting for treatment in many hospitals in East Africa.11 Infants with severe hydrocephalus or hydranencephaly often present with extreme macrocephaly, poor feeding, malnutrition, and scalp ulceration. Their risks associated with ventriculoperitoneal (VP) shunts, particularly the risks of infection, CSF leakage and wound breakdown, are appreciably increased in these patients—to 50% in one series.9

Choroid plexus coagulation (CPC) is one treatment option to manage hydrocephalus and hydranencephaly.12 It has been performed microscopically or endoscopically since the early 1900s, but in recent years has become more popular in combination with endoscopic third ventriculostomy (ETV).5,11 Warf reported that adding CPC to ETV in infants with non-postinfectious hydrocephalus increased the success rate from 38% for ETV alone to 70% for ETV plus CPC.11 However, in many infants with severe hydrocephalus and in those with hydranencephaly, an ETV cannot be done, either because there is no third ventricle or the anatomy is too distorted. We have treated these infants with CPC and report herein our experience with 33 such patients.

Methods

After approval was obtained from the institutional review board of Kijabe Hospital, we conducted a retrospective review of the medical records of infants less than 3 years of age who underwent CPC without ETV from November 2010 to April 2013 and who had undergone no prior treatment of hydrocephalus. Thirty-three infants were identified (Table 1). Follow-up data were obtained
TABLE 1: Summary of clinical and demographic characteristics of 33 patients with extreme hydrocephalus or hydranencephaly treated with CPC

<table>
<thead>
<tr>
<th>Variable</th>
<th>Value</th>
</tr>
</thead>
<tbody>
<tr>
<td>age at surgery</td>
<td></td>
</tr>
<tr>
<td>median</td>
<td>8 wks</td>
</tr>
<tr>
<td>range</td>
<td>0–144 wks</td>
</tr>
<tr>
<td>≤8 wks</td>
<td>17 (51.5)</td>
</tr>
<tr>
<td>9–16 wks</td>
<td>6 (18.2)</td>
</tr>
<tr>
<td>17–24 wks</td>
<td>4 (12.1)</td>
</tr>
<tr>
<td>&gt;24 wks</td>
<td>6 (18.2)</td>
</tr>
<tr>
<td>sex</td>
<td></td>
</tr>
<tr>
<td>male</td>
<td>16 (48.5)</td>
</tr>
<tr>
<td>female</td>
<td>17 (51.5)</td>
</tr>
<tr>
<td>head circumference</td>
<td></td>
</tr>
<tr>
<td>at admission (mean ± SD)</td>
<td>53.06 ± 6.5 cm</td>
</tr>
<tr>
<td>at failure</td>
<td>54.89 cm</td>
</tr>
<tr>
<td>range</td>
<td>43.5–85.3 cm</td>
</tr>
<tr>
<td>diagnosis</td>
<td></td>
</tr>
<tr>
<td>hydranencephaly</td>
<td>11 (33.3)</td>
</tr>
<tr>
<td>severe hydrocephalus</td>
<td>22 (66.7)</td>
</tr>
<tr>
<td>etiology of hydrocephalus/hydranencephaly</td>
<td></td>
</tr>
<tr>
<td>congenital</td>
<td>27 (81.8)</td>
</tr>
<tr>
<td>postinfectious</td>
<td>6 (18.2)</td>
</tr>
</tbody>
</table>

* Values represent numbers of patients (%) unless otherwise indicated.

from outpatient clinic visits, from mobile clinic visits, or by telephone.

Preoperative radiological diagnosis of hydranencephaly was based on ultrasound or CT scan findings of bilateral absence of virtually all supratentorial structures dorsal to the thalami. Infants with severe hydrocephalus retained a thin rim of cortical tissue.

Choroid plexus coagulation was performed with a rigid-lens endoscope (Storz Oi endoscope) inserted through the right anterior fontanel. The choroid plexus was coagulated bilaterally from that approach with a Bugbee wire and monopolar cautery, proceeding posteriorly from the foramen of Monro to the atrium of the ventricles but not into the temporal horns, and in infants with hydranencephaly, all choroid plexus on the dorsum of the thalami was coagulated bilaterally. In infants with severe hydrocephalus, the septum pellucidum was either absent or perforated sufficiently so that bilateral CPC procedures could be done from a unilateral approach. CPC procedures were done without continuous irrigation.

Statistical analyses were performed using SPSS software (standard version 20; SPSS, Inc.). Continuous variables were reported as the mean ± SD or median. Categorical variables were recorded using numbers and percentages. A comparison of the mean values was performed using a paired t-test. Categorical variables were compared using chi-square and Fisher’s exact tests. A probability value of 0.05 was considered significant.

Results

Twenty-two infants had severe hydrocephalus with a minimal cortical mantle; and 11 infants had hydranencephaly. The infants’ head circumferences at admission ranged from 39 to 68.5 cm (mean 53 cm). Their conditions were congenital in 27 cases (81.8%) and postinfectious in 6 (18.2%). One infant had an associated myelomeningocele.

Three patients were lost to follow-up and could not be evaluated; 9 patients were followed in the Kijabe Hospital outpatient clinic, 15 in mobile clinics, and 6 by phone. Of the 6 followed by phone, 2 had a successful procedure and were living asymptotically at home, 2 subsequently underwent VP shunt placement and died 6 months and 10 months thereafter, and 2 died 1 month and 8 months after CPC without having undergone shunt placement.

Choroid plexus coagulation was successful in 13 (43.3%) of the 30 evaluable patients. Of the 20 evaluable infants who were treated with CPC for hydrocephalus, 8 had successful outcomes. Of the 10 evaluable infants with hydranencephaly, 5 had successful outcomes. The most common presenting symptom in the failure group of 17 infants was increased head circumference, which was present in 14 cases, followed by CSF leakage (3 cases). Six patients died after CPC, 2 in the successful outcome group, 3 in the treatment failure group, and 1 while in the hospital after an intraoperative cardiac arrest. CPC failures occurred as early as 8 days and as long as 608 days after the procedure.

Of the 17 patients in whom CPC failed, 10 were subsequently treated with a VP shunt. Of the 7 who were not treated with CSF shunting, 3 were lost to follow-up, 2 received palliative/comfort care, and 1 had an increase in head circumference by 5 cm, which then stabilized with the patient remaining asymptomatic.

Infants who had a head circumference less than 50 cm at admission had a significantly greater likelihood of CPC failure (p = 0.007, OR 17.14, 95% CI 1.79–163.81). The mean head circumference of the CPC failure group enlarged significantly between admission and CPC failure (p = 0.045).

Discussion

Endoscopic coagulation of the choroid plexus was described first by Putnam in 1934. In 1970, Scarff published the first large series of CPC cases, his personal series of 39 children treated during a 23-year period. He reported long-term control of hydrocephalus in 67% of patients. After Milhorat reported in 1974 that CPC in rhesus monkeys reduced CSF production by only 40%, he reported that the use of CPC declined in favor of VP shunts. In 1995, Popple and Ettles reported the results of CPC in 104 patients; their mean age at operation was 2 years and the median 5 months. The authors used an occipital approach and performed bilateral CPC procedures in one sitting for 10 years and unilateral CPC procedures in one sitting thereafter. Of the 80 infants who underwent CPC, 38% did not require a shunt. The success rate was significantly higher in infants with communicating hydrocephalus and in those whose anterior fontanels were not
CPC for hydrocephalus and hydranencephaly
tense. In children with slowly progressive communicating
hydrocephalus without tense fontanels, the success rate was 64%. Major complications developed in 4 children, but there was no death attributable to the surgery. The authors concluded that the main indication for CPC was as the initial treatment of mildly progressive communicates
hydrocephalus in infants.

The success rate for CPC in our series (43.3%) is comparable to that of Poppe and Ettles, and of Sandberg et al., who reported 50% success in 8 patients. Individual practitioners will have to decide whether a success rate of approximately 40% is sufficiently high to avoid the complications of a shunt in children with severe hydrocephalus or hydranencephaly. Those conditions are seen infrequently in developed countries but are common in developing countries, where CPC may be more appropriate because complications of VP shunts are appreciably higher. In a retrospective review of 345 cases involving children with VP shunts at Kenyatta National Hospital, Nairobi, Mwang’ombo et al. found an infection rate of 25%. In Dar es Salaam, Kinasha et al. reported an infection rate of 25%, an obstruction rate of 32%, and a combined complication rate of 46% in 65 children, 18% of whom were less than 6 months old.

Avoidance of a CSF shunt is particularly desirable in infants with extreme macrocephaly. Their risk of shunt complications is increased by the thinness and fragility of their scalp, their usual malnourished condition, and the common presence of infected scalp ulcers at the parietal bosses. For such infants, CPC would seem to be the initial procedure of choice, and was effective in 2 of 3 infants in our initial report.1 The prospective study reported by Malheiros et al. compared CPC with VP shunt placement in 17 infants with either hydranencephaly or near-hydranencephaly.10 CPC successfully controlled head size in 8 of 9 infants. Of 7 children treated with VP shunts, 2 required shunt revisions during follow-up. The authors concluded that endoscopic CPC represents a single, definitive, safe, effective, and economical treatment of hydranencephaly that may avoid the complications of CSF shunting.

Conclusions

We conclude that endoscopic CPC effectively treats severe hydrocephalus and hydranencephaly in approximately 40% of cases and should be considered as an alternative to placement of a VP shunt for infants with these conditions.

Disclosure

The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

Author contributions to the study and manuscript preparation include the following. Conception and design: Albright, Wittayanakorn, Okechi. Acquisition of data: Shitsama, Wittayanakorn, Okechi. Analysis and interpretation of data: Albright, Shitsama, Wittayanakorn. Drafting the article: Shitsama, Wittayanakorn. Critically revising the article: all authors. Reviewed submitted version of manuscript: all authors. Approved the final version of the manuscript on behalf of all authors: Albright. Statistical analysis: Wittayanakorn. Study supervision: Albright.

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