A rachnoid cysts are congenital lesions filled with cerebrospinal fluid (CSF) that do not communicate with the ventricles or the subarachnoid space. These cysts are formed during development through the splitting of arachnoid membranes and can enlarge over time from the entrapment of CSF through a valve-like opening or through the secretion of fluid from the cyst wall. Overall, arachnoid cysts have a prevalence of 1.1%–2.6% in the general population and carry a male/female prevalence ratio of 4:1. Arachnoid cysts are most commonly located in the sylvian fissure (49%), cerebellar hemisphere (24%), occipital lobe (11%), and quadrigeminal cistern (5%–10%).

Quadrigeminal cistern arachnoid cysts (QACs), also described as tectal or pineal region arachnoid cysts, account for 5%–10% of arachnoid cysts. While often asymptomatic and detected incidentally, large QACs can become symptomatic if they compress the brainstem or pineal region or cause obstructive hydrocephalus. Children commonly present with symptoms of hydrocephalus, macrocephaly, developmental delay, or ocular symptoms from pineal compression, but when patients make it...
to adulthood without symptoms, QACs are less likely to progress clinically or radiographically. Asymptomatic arachnoid cysts, especially if stable on serial imaging, are often observed. Indications for surgery include interval growth, development of symptoms from mass effect, and progressive hydrocephalus.

The management of QACs is controversial; surgical options include endoscopic fenestration, open fenestration with or without cyst wall excision, and cystoperitoneal shunting. Treatment of associated hydrocephalus via ventriculoperitoneal (VP) shunting or endoscopic third ventriculostomy (ETV) is often performed concurrently or in a delayed fashion. Given the limited data on the management of QACs, we retrospectively reviewed 20 years of cases managed at our institution and performed a literature review on this topic.

Methods
Institutional Case Series
Retrospective review of the senior author’s personal case logs revealed patients with QACs treated surgically between 2001 and 2021. Patients were excluded if they had been initially treated for their QAC at an outside institution. Preoperative clinic notes, radiologist documentation, intraoperative records, and follow-up clinic notes were reviewed. All patient data remained de-identified through the data extraction process. Statistical analysis was conducted in R (2020, R Foundation for Statistical Computing), and a p value < 0.05 was set as the threshold for statistical significance. The decision of the surgical approach was made by the primary neurosurgeon based on radiographic findings, patient symptomatology, age, and overall clinical picture, with an effort to avoid placement of a shunt whenever possible. Generally, patients with symptomatic or enlarging cysts were treated with endoscopic fenestration (Figs. 1 and 2), and those with symptomatic hydrocephalus were treated with endoscopic fenestration and concurrent ETV (Fig. 3).

Literature Review
We conducted a literature search on PubMed for prior studies describing cases of QACs, with particular attention focused on indications for treatment, symptoms at presentation, treatment modality, follow-up period, outcomes, and complications. Our search terms were as follows: [“arachnoid cyst” OR “arachnoid cysts” AND “tec-
tal” OR “quadrigeminal” OR “quadrigeminal cistern” OR “pineal” AND “fenestration” OR “shunt” OR “shunting” OR “microsurgery” OR “surgery” OR “endoscopy” OR “endoscopic” NOT “spinal”]. We included studies published between 1980 to 2021 reporting at least 5 patients treated for arachnoid cysts located in the quadrigeminal region. We excluded studies with fewer than 5 patients or arachnoid cysts not located in the quadrigeminal region or studies that were an abstract, an editorial, a case report, a review, a natural history study, or translated from another language. Two authors (H.C. and M.A.S.) independently screened abstracts for validation of inclusion criteria and extracted data. Data were collected on the number of patients in each study, indications for treatment, symptoms at presentation, treatment modality, follow-up time, outcome (symptomatic relief and recurrence rate), and complications (morbidity, mortality, and reintervention).

Results
Institutional Case Series
A total of 12 patients with QACs treated at our institution between 2001 and 2021 met our inclusion criteria and were included in our analysis (Table 1). Our cohort consisted of 11 children and 1 adult, aged 1 week to 33 years, with a median age of 9 months. Mean cyst size was 5.1 cm. Hydrocephalus was present in 10 patients (83%).

Endoscopic cyst fenestration was the first intervention in 11 patients (92%), and ETV was performed concurrently in 3 (27%) of these patients. One patient, at age 33 years, was initially treated with ETV because of progressive symptomatic hydrocephalus with minimal mass effect of the cyst; thus, it was determined that the additional risk of cyst fenestration outweighed the benefits (Tables 1 and 2). Reintervention was required in 5 (42%) of 12 patients. One patient required reintervention after an aborted endoscopic fenestration from a coronal approach because of unfavorable anatomy, with brain parenchyma and large vessels overlying the cyst; therefore, it was deemed unsafe to fenestrate from this approach. The patient returned to the operating room several days later, and endoscopic fenestration from a posterior approach was successful. Thus, the need for reoperation after technically successful fenestration from a posterior approach was successful. Two other patients underwent repeat fenestration, one with concurrent cystoperitoneal shunt placement, and a good outcome was achieved in both.

Patients requiring reoperation had a lower median age of 5 months than the patients in whom initial treatment was successful (24.3 months; p = 0.018, Mann-Whitney

FIG. 2. Case 8. This 2-year-old boy presented with headaches and a progressively enlarging QAC (A–C). He did not have significant hydrocephalus and was treated with endoscopic fenestration via a right frontal approach. He tolerated the procedure well, his headaches resolved, and postoperative imaging showed a decrease in the size of the cyst and ventricles (D–F). No CSF diversion was necessary in this case, and endoscopic fenestration alone resolved the patient’s symptoms.
Excluding the 1 adult in the series, the patients who eventually required reoperation still had a statistically significant lower median age at initial intervention ($p = 0.028$, Mann-Whitney U-test). Among the cases that underwent reoperation ($n = 5$), the mean cyst diameter was 5.44 cm compared to the mean diameter of 4.89 cm among cases that did not undergo reintervention ($n = 7$; $p = 0.6687$, 2-sample t-test). Additionally, hydrocephalus at the initial intervention was present in 4 (80%) of the 5 cases that underwent reoperation compared to 6 (85.7%) of the 7 cases that did not undergo reintervention ($p > 0.99$, Fisher’s exact test). Headache was present in 3 cases that did not require reoperation and none of the cases that underwent reintervention ($p = 0.2045$, Fisher’s exact test).

After a mean follow-up of 5.4 years, 83% of patients had improvement or resolution of their symptoms. At the last follow-up, cyst size was decreased in 8 patients (67%), 2 of whom had complete disappearance of the cyst. Aside from the aforementioned need to return to the operating room in 5 patients, there were no major complications. One patient who had undergone initial ETV had a small superficial stitch infection, which resolved with oral antibiotics.

**Literature Review of QACs**

Our literature search identified 94 references published between 1980 and 2021 that met our inclusion criteria based on the screening of abstracts. Of those identified, 87 studies were excluded upon full-text review based on our exclusion criteria, resulting in 7 studies that were included in our analysis (Table 3). These 7 studies reported a total of 108 patients with QACs with a mean age of 8.8 years. Eighty-seven percent ($n = 94$) of patients had hydrocephalus, and 90% ($n = 97$) were symptomatic at the time of presentation. The mean follow-up duration was 3.4 years.

The majority of cases were treated with endoscopic fenestration as the first intervention (92%, $n = 99$). ETV alone was used as the first intervention in 2% of cases ($n = 2$), craniotomy for excision represented the first intervention in 3% of cases ($n = 3$), and VP shunting was used in 4% of cases ($n = 4$). Of those treated with endoscopic fenestration as the first intervention, concurrent ETV was performed in 44% ($n = 48$). Overall, cyst reduction occurred in 77% of cases ($n = 83$) and symptomatic resolution in 68% of cases ($n = 73$). Of those treated with endoscopic fenestration as the primary intervention, cyst reduction occurred in 79% of cases ($n = 78$) and symptomatic cure in 69% of cases ($n = 68$). Complete radiographic resolution occurred in none of the cases (Tables 2 and 3).

Complications occurred in 18% of cases ($n = 19$), including 16% of endoscopic fenestration cases ($n = 16$). Complications included subdural collections ($n = 12$), intraoperative bleeding ($n = 3$), pseudomeningocele ($n = 2$), air embolism ($n = 1$), and CSF leakage ($n = 1$). Reintervention occurred in 31% of all cases ($n = 33$) and 29% of the cases initially treated with endoscopic fenestration.
<table>
<thead>
<tr>
<th>Case No.</th>
<th>Sex</th>
<th>Max Cyst Diameter (cm)</th>
<th>Presenting Sxs</th>
<th>Hydrocephalus at Presentation</th>
<th>Age at Tx</th>
<th>Initial Tx</th>
<th>No. of Subsequent Txs</th>
<th>2nd Tx</th>
<th>3rd Tx</th>
<th>4th Tx</th>
<th>FU (yrs)</th>
<th>Sxs at Last FU</th>
<th>Cyst Size at Last FU</th>
<th>Ventricle Size at Last FU</th>
<th>Sxs at Last FU</th>
<th>Cyst Size at Last FU</th>
<th>Ventricle Size at Last FU</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>F</td>
<td>2.5</td>
<td>Macro</td>
<td>Yes</td>
<td>2 yrs</td>
<td>EF</td>
<td>0</td>
<td>0</td>
<td></td>
<td></td>
<td>12</td>
<td>Improved</td>
<td>No residual</td>
<td>Mod</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>2</td>
<td>M</td>
<td>7</td>
<td>Macro</td>
<td>Yes</td>
<td>1 wk</td>
<td>EF</td>
<td>0</td>
<td>1</td>
<td>EF, CP shunt</td>
<td></td>
<td>9</td>
<td>Resolved</td>
<td>Same</td>
<td>Mod</td>
<td></td>
<td></td>
<td></td>
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<td>3</td>
<td>F</td>
<td>5</td>
<td>Incidental</td>
<td>No</td>
<td>5 mos</td>
<td>EF</td>
<td>0</td>
<td>1</td>
<td>EF</td>
<td></td>
<td>1.5</td>
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<td>Same</td>
<td>Dilated</td>
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<td>4</td>
<td>M</td>
<td>5.8</td>
<td>Micro</td>
<td>Yes</td>
<td>4 mos</td>
<td>EF</td>
<td>0</td>
<td>6</td>
<td>VP shunt</td>
<td>CP shunt</td>
<td>13</td>
<td>Resolved</td>
<td>No residual</td>
<td>Mod</td>
<td></td>
<td></td>
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<tr>
<td>5</td>
<td>F</td>
<td>6.5</td>
<td>Incidental</td>
<td>Yes</td>
<td>10 mos</td>
<td>EF, ET V</td>
<td>0</td>
<td>0</td>
<td></td>
<td></td>
<td>9</td>
<td>Resolved</td>
<td>Decreased</td>
<td>Dilated</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>6</td>
<td>F</td>
<td>3.2</td>
<td>Macro</td>
<td>Yes</td>
<td>8 mos</td>
<td>EF, ET V</td>
<td>0</td>
<td>1</td>
<td>Craniotomy for cyst excision</td>
<td></td>
<td>7</td>
<td>Resolved</td>
<td>Same</td>
<td>Small</td>
<td></td>
<td></td>
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<td>7</td>
<td>M</td>
<td>3.4</td>
<td>HA, gait difficulty</td>
<td>Yes</td>
<td>19 yrs</td>
<td>EF</td>
<td>0</td>
<td>0</td>
<td></td>
<td></td>
<td>5*</td>
<td>Improved</td>
<td>Decreased</td>
<td>Mod</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>8</td>
<td>M</td>
<td>5.6</td>
<td>HA</td>
<td>No</td>
<td>2 yrs</td>
<td>EF</td>
<td>0</td>
<td>0</td>
<td></td>
<td></td>
<td>5</td>
<td>Resolved</td>
<td>Decreased</td>
<td>Mod</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>9</td>
<td>F</td>
<td>1.5</td>
<td>HA</td>
<td>Yes</td>
<td>33 yrs</td>
<td>ET V</td>
<td>Superficial stitch infection resolved w/ antibiotics</td>
<td>0</td>
<td></td>
<td></td>
<td>2*</td>
<td>Resolved</td>
<td>Decreased</td>
<td>Mod</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>10</td>
<td>M</td>
<td>8.6</td>
<td>Macro, DG</td>
<td>Yes</td>
<td>5 mos</td>
<td>EF, septostomy</td>
<td>0</td>
<td>0</td>
<td></td>
<td></td>
<td>4</td>
<td>Improved</td>
<td>Decreased</td>
<td>Mod</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>11</td>
<td>M</td>
<td>6.2</td>
<td>Macro</td>
<td>Yes</td>
<td>6 mos</td>
<td>EF (attempted)</td>
<td>Surgery aborted</td>
<td>1</td>
<td>EF</td>
<td></td>
<td>2</td>
<td>Resolved</td>
<td>Same</td>
<td>Mod</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>12</td>
<td>M</td>
<td>6.1</td>
<td>Macro</td>
<td>Yes</td>
<td>7 yrs</td>
<td>EF, ET V</td>
<td>0</td>
<td>0</td>
<td></td>
<td></td>
<td>2</td>
<td>Same</td>
<td>Decreased</td>
<td>Mod</td>
<td></td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

CP = cystoperitoneal; Cx = complication; DG = dysconjugate gaze; EF = endoscopic fenestration; FU = follow-up; HA = headache; macro = macrocephaly; micro = microcephaly; mod = moderate; Sx = symptom; Tx = treatment.

* Months.
(n = 29). Reinterventions included VP shunt placement (n = 15), repeat fenestration (n = 13), ETV (n = 1), cyst wall excision (n = 1), and cystoperitoneal shunt placement (n = 3). Of the repeat fenestration cases, 3 were performed with the addition of ETV and 5 with VP shunting. One case that underwent repeat fenestration with ETV required a third intervention (an additional re-fenestration and ETV). Two of the 3 cases that were treated initially with craniotomy required reintervention (1 ETV, 1 VP shunt placement).

**Discussion**

Optimal management of QACs is controversial, as these patients frequently require reintervention. Additionally, the high rate of concurrent hydrocephalus introduces added complexity to surgical decision-making. A retrospective review of 20 years of patients treated at our institution and a review of the literature revealed several nuances. To the best of our knowledge, this is the first reported comprehensive literature review of QACs.

Retrospective analysis of patients treated at our institution revealed that after technically successful endoscopic fenestration, 33% of patients required reintervention because of progressive symptoms. A literature review similarly revealed that 29% of patients who had undergone initial endoscopic fenestration required reintervention. Some authors have suggested that early intervention for arachnoid cysts is important for proper brain development, however, our data suggested that younger patients are at higher risk for requiring repeat surgery. We observed a statistically significant association with a younger age in the group of patients who required reintervention (median 5 months) compared to the age of patients who did not (median 24.3 months). There were no statistically significant differences between the two groups regarding cyst diameter, presence of hydrocephalus, or headache at initial presentation. Although our retrospective case series is limited by a small sample size, our literature review demonstrated that the 3 studies with the youngest average age had the three highest rates of reintervention. However, additional data are needed to corroborate our findings.

Some authors have suggested a need to treat the hydrocephalus concurrently with cyst fenestration, and some groups routinely perform ETV concurrently with endoscopic fenestration in most cases. Studies by Cinalli et al. and Erşahin and Kesiği revealed that shunt independence and fenestration success were associated with the addition of ETV when performing cyst fenestration. In the study from Cinalli et al., 7 of the 8 cases treated with ETV without fenestration without fenestration had undergone concurrent ETV required re-operation with ETV.

Our practice, and that of others, has been to identify the source of the patient’s symptoms, whether from mass effect due to the cyst and/or from hydrocephalus, and to tailor the treatment accordingly, with the goal of avoiding permanent shunt placement whenever possible. Generally, patients with progressive symptoms from the cyst are treated with endoscopic fenestration, and cases with concurrent symptomatic hydrocephalus are treated with concurrent ETV (Fig. 1). In our retrospective cohort, only 3 (27%) of the 11 patients initially treated with endoscopic fenestration underwent concurrent ETV, and 1 (25%) of 4 patients who required reintervention after failed fenestration had undergone concurrent ETV. Our data suggested that upfront treatment of hydrocephalus is not an independent predictor of the need for reintervention. Careful selection of patients who require direct treatment of their hydrocephalus appears to be more important. Indeed, the literature revealed that 46% of reintervention procedures were for VP shunting (whereas just 39% underwent re-fenestration). When also considering patients who underwent combined re-fenestration and treatment of hydrocephalus, a total of 73% of reintervention procedures involved the treatment of hydrocephalus. This finding suggests that the most frequent reason for reintervention is untreated or unresolved hydrocephalus after the first procedure, and identifying patients who require initial treatment of hydrocephalus is critically important.

**Conclusions**

QACs that come to surgery will frequently require reintervention. Endoscopic fenestration appears to be safe and effective for these lesions, and treatment of associated hydrocephalus by ETV or VP shunting may be necessary.
<table>
<thead>
<tr>
<th>Authors &amp; Year</th>
<th>Years of Study</th>
<th>Total No. of Patients</th>
<th>Mean Patient Age (yrs)</th>
<th>% w/ Hydrocephalus</th>
<th>% Symptomatic</th>
<th>Initial Tx</th>
<th>Symptomatic Cure</th>
<th>Reintervention</th>
<th>Cxs</th>
<th>Cyst Reduction</th>
<th>Mean FU (yrs)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Yu et al., 2016&lt;sup&gt;16&lt;/sup&gt;</td>
<td>2007–2013</td>
<td>8</td>
<td>23</td>
<td>50%</td>
<td>100%</td>
<td>EF (8/8, 100%; &amp; ETV 3/8, 38%)</td>
<td>5 (63%)</td>
<td>1 (13%): re-fenestration</td>
<td>NR</td>
<td>5 (63%)</td>
<td>4.28</td>
</tr>
<tr>
<td>Gui et al., 2016&lt;sup&gt;17&lt;/sup&gt;</td>
<td>2007–2014</td>
<td>28</td>
<td>8.2</td>
<td>100%</td>
<td>100%</td>
<td>EF (28/28, 100%)</td>
<td>25 (89%)</td>
<td>3 (11%): 1 re-fenestration, 2 VP shunt</td>
<td>4 (14%): subdural collections</td>
<td>22 (79%)</td>
<td>4.06</td>
</tr>
<tr>
<td>Garg et al., 2014&lt;sup&gt;18&lt;/sup&gt;</td>
<td>2002–2012</td>
<td>18</td>
<td>17</td>
<td>100%</td>
<td>100%</td>
<td>EF (9/18, 50%; &amp; ETV 7/18, 39%), craniotomy for excision &amp; fenestration (3/18, 17%), VP shunt (4/18, 22%), ETV (2/18, 11%)</td>
<td>11 (61%)</td>
<td>7 (39%): 1 ETV, 1 excision, 5 VP shunt</td>
<td>5 (28%): pseudomeningocele (2), air embolism during sitting craniotomy (1), intraop endoscopy bleed (1), SDH (1)</td>
<td>12 (67%)</td>
<td>1.98</td>
</tr>
<tr>
<td>Gangemi et al., 2005&lt;sup&gt;19&lt;/sup&gt;</td>
<td>1993–2004</td>
<td>5</td>
<td>5.5</td>
<td>100%</td>
<td>100%</td>
<td>EF &amp; ETV (5/5, 100%)</td>
<td>4 (80%)</td>
<td>1 (20%): CP shunt</td>
<td>NR</td>
<td>3 (60%)</td>
<td>0.42</td>
</tr>
<tr>
<td>Cinalli et al., 2010&lt;sup&gt;20&lt;/sup&gt;</td>
<td>1995–2008</td>
<td>14</td>
<td>2.2</td>
<td>64%</td>
<td>79%</td>
<td>EF (14/14, 100%; &amp; ETV 6/14, 43%)</td>
<td>7 (50%)</td>
<td>7 (50%): 1 re-fenestration, 3 re-fenestration &amp; ETV (2x in 1 of these patients), 1 VP shunt, 2 CP shunt</td>
<td>3 (21%): subdural hemorrhage (1), intraop bleeding &amp; postop CSF leak (1), intraop bleeding (1)</td>
<td>11 (79%)</td>
<td>4.58</td>
</tr>
<tr>
<td>El-Ghandour, 2013&lt;sup&gt;21&lt;/sup&gt;</td>
<td>2003–2011</td>
<td>18</td>
<td>2.5</td>
<td>100%</td>
<td>100%</td>
<td>EF (18/18, 100%; &amp; ETV 14/18, 78%)</td>
<td>11 (61%)</td>
<td>7 (39%): 2 re-fenestration, 5 re-fenestration &amp; VP shunt</td>
<td>3 (17%): subdural hygroma</td>
<td>14 (78%)</td>
<td>3.82</td>
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<td>Ergahin &amp; Kesikçi, 2009&lt;sup&gt;22&lt;/sup&gt;</td>
<td>2000–2007</td>
<td>17</td>
<td>3.3</td>
<td>100%</td>
<td>100%</td>
<td>EF (17/17, 100%; &amp; ETV 13/17, 76%)</td>
<td>10 (59%)</td>
<td>7 (41%): 7 VP shunt</td>
<td>4 (24%): subdural fluid collection (3), CSF fistula leak &amp; meningitis (1)</td>
<td>16 (94%)</td>
<td>4.32</td>
</tr>
</tbody>
</table>

NR = none reported; SDH = subdural hematoma.
Younger patients may be at higher risk for reintervention, and the literature suggests that untreated hydrocephalus is a common cause of reintervention.

References
15. Raffel C, McComb JG. To shunt or to fenestrate: which is the best surgical treatment for arachnoid cysts in pediatric patients? Neurosurgery. 1988;23(3):338-342.

Disclosures
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
Conception and design: Silva. Acquisition of data: Silva, Chang, Weng, Hernandez, Ragheb. Analysis and interpretation of data: all authors. Drafting the article: Silva, Chang. Critically revising the article: Silva, Chang, Shah, Wang, Niazi, Ragheb. Reviewed submitted version of manuscript: Silva, Chang, Shah, Wang, Niazi, Ragheb. Approved the final version of the manuscript on behalf of all authors: Silva. Statistical analysis: Silva, Chang. Administrative/technical/material support: Silva, Weng, Hernandez, Ragheb. Study supervision: Silva, Ragheb.

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