Natural history of Chiari malformation Type I following decision for conservative treatment

Clinical article

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Object: The natural history of the Chiari malformation Type I (CM-I) is incompletely understood. The authors report on the outcome of a large group of patients with CM-I that were initially selected for nonsurgical management.

Methods: The authors identified 147 patients in whom CM-I was diagnosed on MR imaging, who were not offered surgery at the time of diagnosis, and in whom at least 1 year of clinical and MR imaging follow-up was available after the initial CM-I diagnosis. These patients were included in an outcome analysis.

Results: Patients were followed clinically and by MR imaging for a mean duration of 4.6 and 3.8 years, respectively. Of the 147 patients, 9 had new symptoms attributed to the CM-I during the follow-up interval. During this time, development of a spinal cord syrinx occurred in 8 patients; 5 of these patients had a prior diagnosis of a presyrinx state or a dilated central canal. Spontaneous resolution of a syrinx occurred in 3 patients. Multiple CSF flow studies were obtained in 74 patients. Of these patients, 23 had improvement in CSF flow, 39 had no change, and 12 showed worsening CSF flow at the foramen magnum. There was no significant change in the mean amount of cerebellar tonsillar herniation over the follow-up period. Fourteen patients underwent surgical treatment for CM-I. There were no differences in initial cerebellar tonsillar herniation or CSF flow at the foramen magnum in those who ultimately underwent surgery compared with those who did not.

Conclusions: In patients with CM-I that are selected for nonsurgical management, the natural history is usually benign, although spontaneous improvement and worsening are occasionally seen. (DOI: 10.3171/2011.5.PEDS1122)

Key Words • Chiari malformation • syrinx • natural history

Children with Chiari malformation Type I (CM-I) discovered on MR imaging are frequently referred for neurosurgical evaluation. Most reported series of CM-Is describe outcomes for patients who have been selected for surgery. As a consequence, the natural history of CM-Is is poorly understood. Recently, several groups have reported on the natural history of CM-Is in a small number of patients in whom the entity was managed without surgery. Novegno et al. reported on 22 children with CM-I, 17 of whom remained asymptomatic or improved over follow-up intervals of 3–19 years. Only 1 patient in that series had an associated spinal cord syrinx. Nishizawa et al. reported on 9 adults with incidentally discovered asymptomatic CM-I and syrinx that were managed without surgery, with a mean follow-up interval of 11 years. Of these patients, 8 remained clinically stable and 1 patient was treated surgically for new neurological symptoms. These small case series suggest a relatively benign natural history for asymptomatic or minimally symptomatic CM-I.

There are no universally accepted criteria for selecting patients with CM-I for surgical versus nonsurgical treatment. Haines and Berger surveyed pediatric neurosurgeons in 1991 and found a wide variety of opinions on the indications for surgical treatment. They suggested that at that time a natural history analysis of asymptomatic cases would be useful. More recent surveys of pediatric neurosurgeons have continued to show differences of opinion regarding surgical indications for CM-I. Schijman and Steinbok surveyed pediatric neurosurgeons on CM-I management in 2004. That survey found that 8% of pediatric neurosurgeons would recommend surgical treatment for an asymptomatic patient with CM-I, and that 75% would recommend surgical treatment if a spinal cord syrinx was present. This tendency to treat
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CM-I surgically if a syrinx is present has made natural history analysis of this subgroup particularly challenging. We hope that an analysis of a large group of patients with CM-I with or without associated spinal cord syrinx that have been selected for nonsurgical management will improve our understanding of the natural history of these conditions, and ultimately improve surgical management decisions for this group of patients.

Methods

Medical Records Search

Following approval by the Institutional Review Board at the University of Michigan, we performed a retrospective review of the electronic medical records of all patients ≤ 18 years of age who underwent brain or cervical spine MR imaging at the University of Michigan between November 1997 and August 2008. Brain or cervical spine MR images were obtained in 14,116 individual children during this period. All MR images were performed on either a 1.5- or 3-T MR imaging device. Electronic records were reviewed using the Electronic Medical Record Search Engine (EMERSE), a search engine that queries all free-text documents within the electronic medical records of a specified patient population. In this study, we used EMERSE to identify a population in whom the terms “tonsillar ectopia,” “tonsillar herniation,” “tonsillar descent,” “syrinx,” “syringomyelia,” “hydromyelia,” and “Chiari” were used in any part of the medical record. We then reviewed the records of all patients identified by EMERSE to select those who met the criteria for inclusion.

All included patients had cerebellar tonsillar ectopia measured as ≥ 5 mm below the foramen magnum. Also, to be included in the analysis, all patients who were treated nonsurgically must have had at least a 1-year interval between the first and last clinical assessment and at least a 1-year interval between the first and last MR imaging session. Any patient who underwent surgical treatment for a CM-I on the basis of a recommendation made at the initial neurosurgical consultation was excluded from analysis. Patients who were treated surgically at an interval following an initial recommendation for nonsurgical management were included, even if the interval between the initial evaluation and surgical treatment was < 1 year. Patients were excluded if the cerebellar tonsillar descent was believed to be secondary to mass effect from other intracranial or cranial conditions such as tumor, cerebral edema, arachnoid cyst, or craniosynostosis. Any patient who met criteria for CM-II, -III, or -IV, including a history of myelomeningocele repair, was excluded from analysis. Finally, any patient who had undergone surgical treatment for CM-I prior to MR imaging evaluation at our institution was excluded.

We identified 509 patients who met the initial inclusion criteria for a diagnosis of CM-I on MR imaging at our institution. Of these, 120 patients were excluded because a recommendation for surgical treatment was made at the first clinical assessment. Another 242 patients were excluded because they had < 1 year of clinical and MR imaging follow-up. A total of 147 patients met all inclusion criteria for the natural history analysis. For these patients, we recorded age and sex, neurological symptoms, other clinical diagnoses, other radiological diagnoses, the indication that was provided for performing the initial MR imaging, and any surgical treatment. In addition, we recorded imaging characteristics of the CM-I, including the amount of cerebellar tonsillar descent, CSF flow at the foramen magnum, and the presence of any spinal cord syrinx. If a patient had more than 2 MR imaging studies, we considered only the studies performed at the time of CM-I diagnosis as well as the most recent ones in nonsurgically treated patients, or the MR imaging studies performed immediately preceding surgery in surgically treated patients. Not all patients underwent complete spine imaging at the time of CM-I diagnosis; therefore, for those patients with a spinal cord syrinx, the initial MR imaging study that demonstrated a syrinx was considered to be the initial spine study for these patients. The CSF flow was categorized as normal, decreased anteriorly or posteriorly at the foramen magnum, or decreased at the foramen magnum with abnormal tonsillar pulsations.

For the purpose of this analysis, the presence of a syrinx was defined as a spinal cord cyst (hypointensity on T1-weighted images, with corresponding T2 hyperintensity) ≥ 3 mm in maximal anteroposterior diameter on sagittal or axial imaging. Presyrinx states (T2 hyperintensity, indistinct T1 prolongation, without cavitation) were classified separately. If a syrinx was present, we recorded its diameter in millimeters at the widest diameter as viewed on sagittal imaging. The length of the syrinx was recorded according to the number of corresponding vertebral levels.

Statistical Analysis

Statistical significance calculations were obtained using ANOVA, the chi-square test, and Tukey multiple comparisons. Univariate logistic regression was used to evaluate change over time. Data were analyzed using SPSS version 16.0 software (SPSS, Inc.).

Results

Patient Population

Included in the analysis were 147 patients ≤ 18 years of age (mean age 7.7 years). The mean duration of MR imaging follow-up was 3.8 years. The mean length of clinical follow-up by a neurosurgeon or neurologist was 4.6 years, and the mean duration of clinical follow-up by any physician at our hospital was 6.5 years. In the study population, 76 (51.7%) patients were female. Other clinical conditions and imaging findings diagnosed in the study group are summarized in Table 1.

Tonsillar Descent

There was no change in mean cerebellar tonsillar herniation for the group as a whole over a mean follow-up of 3.8 years. The mean tonsillar herniation was 9.53 mm at presentation and 9.34 mm at follow-up (p = 0.4). Considered individually, however, many patients did have a change in the amount of tonsillar descent (Fig. 1).
Changes in tonsillar herniation ≤ 2 mm over the follow-up interval were seen in 103 patients (73%). Interval improvement in the amount of tonsillar descent was seen in 45 patients (31%), and 7 patients had a follow-up MR imaging study with < 5 mm tonsillar descent, and were therefore no longer considered to have a CM-I by our criteria. An increase in tonsillar herniation of at least 4 mm was seen in 6 patients (4%).

We attempted to identify factors that were predictive of a change in the degree of tonsillar descent. Sex was not predictive of change in tonsillar descent (p = 0.54). Advancing age was associated (p = 0.007) with a decrease in the amount of tonsillar herniation (regression slope estimate –0.15) (Fig. 2). Patients between 0 and 6 years of age at the time of CM-I diagnosis had a mean increase in tonsillar herniation of 0.63 mm. In contrast, patients between 6 and 12 years of age at the time of CM-I diagnosis had a mean decrease in tonsillar herniation of 0.53 mm, and those between 12 and 18 years of age had a mean decrease in tonsillar herniation of 1.24 mm (p = 0.02). Of the 14 patients who were surgically treated, 11 had interval MR imaging studies prior to surgery. Those patients who were not treated surgically over the follow-up interval had a mean decrease in tonsillar herniation of 0.32 mm, and those treated with surgical decompression during the follow-up interval had a mean increase in tonsillar herniation of 0.93 mm prior to surgery.

Evaluation of CSF Flow

The CSF flow at the foramen magnum was evaluated on MR images obtained in 74 patients at the time of CM-I diagnosis as well as at the time of the most recent follow-up MR imaging session or the MR imaging studies performed prior to surgery. Interval improvement in CSF flow was seen in 23 patients, 39 patients had no change in CSF flow, and 12 patients had decreased CSF flow on the follow-up study compared with the initial MR imaging. The patient’s age at time of CM-I diagnosis was not predictive of changes in CSF flow over the follow-up interval. Of the 14 patients who were treated surgically, 7 had interval CSF flow studies for comparison, and all 7 of these patients had stable (5 patients) or improved (2 patients) CSF flow. Of the 12 patients with decreased CSF flow on follow-up imaging, none had worse symptoms or a new syrinx, and none were treated surgically. A change in CSF flow analysis was never used as a surgical indication in this series of patients.

Syrinx Characteristics

A spinal cord syrinx of at least 3 mm diameter was predictive of change in tonsillar descent (p = 0.54). Advancing age was associated (p = 0.007) with a decrease in the amount of tonsillar herniation (regression slope estimate –0.15) (Fig. 2). Patients between 0 and 6 years of age at the time of CM-I diagnosis had a mean increase in tonsillar herniation of 0.63 mm. In contrast, patients between 6 and 12 years of age at the time of CM-I diagnosis had a mean decrease in tonsillar herniation of 0.53 mm, and those between 12 and 18 years of age had a mean decrease in tonsillar herniation of 1.24 mm (p = 0.02). Of the 14 patients who were surgically treated, 11 had interval MR imaging studies prior to surgery. Those patients who were not treated surgically over the follow-up interval had a mean decrease in tonsillar herniation of 0.32 mm, and those treated with surgical decompression during the follow-up interval had a mean increase in tonsillar herniation of 0.93 mm prior to surgery.

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Syrinx Characteristics

A spinal cord syrinx of at least 3 mm diameter was
found in 20 patients (13.5%). In 13 of these patients, the syrinx was present at the time of CM-I diagnosis. Development of a syrinx during the follow-up interval occurred in 7 patients, with a mean time to syrinx development of 28 months (range 2–74 months). One patient with a thoracic syrinx developed a new cervical spine syrinx during follow-up. Of the 7 new syringes, 2 developed from a previously identified presyrinx state (hyperintensity on T2-weighted MR imaging studies without cavitation), 3 developed from what had been considered a dilated central canal < 3 mm in diameter, and 2 patients had normal results on prior spine MR images (Fig. 3). In the 13 patients with a syrinx at the time of CM-I diagnosis, 6 lesions were unchanged in size, 5 were smaller, and 2 were larger on follow-up MR imaging. Of the 5 patients with spontaneous improvement in syrinx size, 3 demonstrated complete syrinx resolution on follow-up MR imaging.

Despite these examples of individual changes in syrinx dimensions, when the group was analyzed as a whole there was no change in average initial (4 vertebral levels) and final syrinx length (p = 0.88; Fig. 4 left). In addition, there was no significant change in the mean initial (4.25 mm) and final (3.56 mm) syrinx width (p = 0.16; Fig. 4 right). All 5 patients who had a decrease in syrinx size were female. Patients with syrinx progression (mean age 6.7 years) or regression (mean age 5.6 years) were younger (p = 0.05) compared with those whose syrinx remained stable (mean age 11.6 years). Patients with new syrinx formation over the follow-up interval had a mean initial tonsillar herniation of 13.5 mm. Those patients with larger syringes on follow-up imaging had a greater mean initial tonsillar herniation (14.5 mm) compared with those with a stable (8.6 mm) or decreased (8.6 mm) syrinx size (p = 0.04).

Of the 20 patients with a spinal cord syrinx, 6 were treated surgically during the follow-up interval. Of these 6, concern for the syrinx was documented as a primary
consideration in the decision to recommend surgery in 4 cases. The remaining 2 patients underwent surgery for reasons thought to be unrelated to changes in the syrinx. There was an association between eventual surgical treatment and a prior increase in syrinx width (0.7 mm, p = 0.05) and length (2.2 levels, p = 0.08).

**Symptoms of CM-I**

A patient was designated as being symptomatic from their CM-I if either the primary neurologist or neurosurgeon treating the individual at our institution thought that the complement of presenting symptoms was attributable to the lesion. There was no significant difference in the number of symptomatic patients between the time of initial presentation and the most recent follow-up. There were 5 patients who remained symptomatic throughout the follow-up interval. The 6 patients who were symptomatic at presentation were not so at last follow-up. Another 9 patients who had symptoms that were initially not thought to be due to the CM-I were considered to be symptomatic at the time of the most recent evaluation.

**Rationale for Surgery**

Of the 147 patients in the study group, 14 underwent surgery for CM-I (Table 2). The most common reasons for surgical treatment during the follow-up interval were medically refractory and persistent headaches, sleep apnea, and changes in a syrinx. For these 14 patients, the mean time to surgery after CM-I diagnosis was 2.1 years.

**TABLE 2: Results in 14 patients receiving surgical treatment for CM-I following an initial decision for conservative management**

<table>
<thead>
<tr>
<th>Case No.</th>
<th>Preop Symptoms</th>
<th>Time to Op (days)</th>
<th>Descent (mm)</th>
<th>Syrinx</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>syrinx progression</td>
<td>179</td>
<td>8.0</td>
<td>ND yes; new</td>
</tr>
<tr>
<td>2</td>
<td>worsening HAs</td>
<td>183</td>
<td>20.0</td>
<td>20.0 no</td>
</tr>
<tr>
<td>3</td>
<td>sleep apnea</td>
<td>214</td>
<td>16.0</td>
<td>16.0 no</td>
</tr>
<tr>
<td>4</td>
<td>sleep apnea</td>
<td>232</td>
<td>15.0</td>
<td>20.0 no</td>
</tr>
<tr>
<td>5</td>
<td>worsening HAs</td>
<td>367</td>
<td>16.0</td>
<td>ND no</td>
</tr>
<tr>
<td>6</td>
<td>HA refractory to medical therapy</td>
<td>434</td>
<td>15.0</td>
<td>ND no</td>
</tr>
<tr>
<td>7</td>
<td>rapid progression of scoliosis</td>
<td>474</td>
<td>8.0</td>
<td>8.0 yes; new</td>
</tr>
<tr>
<td>8</td>
<td>central sleep apnea</td>
<td>526</td>
<td>10.0</td>
<td>10.0 yes</td>
</tr>
<tr>
<td>9</td>
<td>new syrinx</td>
<td>812</td>
<td>17.0</td>
<td>20.0 yes; new</td>
</tr>
<tr>
<td>10</td>
<td>concern for neurological decline</td>
<td>849</td>
<td>12.0</td>
<td>15.0 yes</td>
</tr>
<tr>
<td>11</td>
<td>concern for neurological decline</td>
<td>1152</td>
<td>12.0</td>
<td>9.0 no</td>
</tr>
<tr>
<td>12</td>
<td>worsening HAs, occipital neck pain</td>
<td>1316</td>
<td>5.5</td>
<td>5.5 no</td>
</tr>
<tr>
<td>13</td>
<td>sleep apnea</td>
<td>1744</td>
<td>12.8</td>
<td>16.0 no</td>
</tr>
<tr>
<td>14</td>
<td>new syrinx</td>
<td>2449</td>
<td>30.0</td>
<td>29.0 yes; new</td>
</tr>
</tbody>
</table>

* For the 3 patients marked “ND,” no interval brain MRI was obtained prior to surgical treatment. Abbreviations: HA = headache; ND = not done.

There was no significant difference in the initial tonsillar herniation in the group that ultimately underwent surgery compared with those individuals who did not undergo surgery. In addition, there were no significant differences in the change in CSF flow at the foramen magnum between the group that underwent surgery and the group that did not have surgery.

**Discussion**

We analyzed the outcome of a group of 147 children with CM-I that was initially managed without surgery. Of these, 14 patients eventually underwent surgical intervention, and 133 patients remained asymptomatic or minimally symptomatic. Spontaneous clinical and radiological improvement was seen in some cases. These data support a generally benign natural history for those patients with CM-I that meet the usual criteria for conservative management.

There is very little existing information on the natural history of CM-I. Novegno et al. recently reported on a series of 22 patients for whom nonsurgical management was recommended. Over a mean follow-up interval of 5.9 years, they found that 5 patients had symptomatic worsening, and 3 of these required surgery. Spontaneous improvement was seen in 3 of their patients, and 1 patient had complete resolution. They concluded that a conservative approach to asymptomatic or minimally symptomatic patients could be justified based on their data. Nishizawa et al. reported on 9 adults with CM-I and a spinal cord syrinx in whom the entity was managed without surgery, and found that 8 of these patients had a benign natural course. There are several reported cases of CM-I with spontaneous improvement in cerebellar tonsillar herniation. In many of these cases, improvement in the degree of tonsillar herniation was associated with a decrease in the size or resolution of the associated syrinx. Because the presence of CM-I on imaging does not have any pathological consequences in many cases, several surgeons have recently suggested a preference for the term “Chiari Type I anomaly” rather than “Chiari malformation.” This suggested change in
Natural history of Chiari malformation Type I
terminology may help to emphasize the generally benign
natural history in many cases.

The pathophysiologic mechanism underlying the
relationship between CM-I and spinal cord syrinx is
well established,2,4,6,8,11,16,17,22,24,26,27,30,32,40 The presence of
a syrinx is an indication for surgical treatment of CM-I
at many centers,13,15,34 and therefore there are few reports
on the natural history of syrinx in the setting of CM-I.5,28
Schijman and Steinbok34 conducted a survey regarding
management of CM-I with and without an associated syr-
inx. Of the responders, 75% stated that they would per-
form a suboccipital decompression for an asymptomatic
patient with CM-I and a syrinx that was at least 8 mm in
maximum diameter, and 28% would intervene in a simi-
lar patient with syrinx that was 2 mm in diameter.

Although it has generally been our practice to rec-
ommend surgical treatment for a CM-I associated with a
spinal syrinx, we identified 13 patients in our series who
were known to have a spinal cord syrinx at the time of
CM-I diagnosis, and whose syringes were managed with-
out surgery. Furthermore, 7 patients without a syrinx at the
time of CM-I diagnosis developed one over the follow-up
interval. In 2 of these cases, the syringes developed from a
presyrinx state, defined as indistinct T2 prolongation with-
out spinal cord cavitation.9,10,21 In our study, 3 additional
patients were found to have syringes that developed from
what had been considered a dilated central canal. One of
these patients was later found to have a decrease in the
width of the syrinx, no longer meeting the definition for a
syrinx in this study.

In our series, the presence of a syrinx was not always
associated with progression of symptoms. Only 6 of 20
patients with a syrinx had surgery during the follow-up
interval, and 2 of these underwent surgery for reasons
other than the syrinx. Other series have also suggested
that surgery may not be necessary in all cases of CM-I
and syrinx.5,28 Nishizawa et al.28 reported on 9 adult pa-
tients with incidental CM-I and syrinx, only 1 of whom
required surgery over a 10-year follow-up interval. They
reported no significant change in the MR imaging char-
acteristics for 8 of their 9 patients over the follow-up in-
terval. In addition to reports showing stability of syringes
over time, there have been cases of regression and com-
plete resolution of syringes.37–39 Sudo and colleagues35,36
were the first to report spontaneous resolution of syrinx
associated with CM-I. Kyoshima and Bogdanov20 report-
ed on 2 cases of cranial-cervical junction abnormalities
with associated syrinx in which spontaneous clinical and
radiographic improvement was seen.

Using the need for surgical treatment as a primary end
point in the evaluation of CM-I natural history has the po-
tential to overestimate the true rate of clinical worsening.
Several of our patients who went on to have surgery after
the initial recommendation for conservative management
had persistent symptoms rather than the onset of new
ones. New symptoms were uncommon in this group of
patients, and often the presence of new symptoms did not
correlate with a decision to pursue surgery. Chiari symp-
toms can be notoriously protean, and may overlap with
other neurological conditions. Any analysis of CM-I that
seeks to categorize patient symptoms according to their
cause will be subjective, especially when viewed retro-
spectively. Furthermore, the presence of symptoms does
not always correlate with the perceived need for surgical
management. Occasionally, patients and surgeons may elect
to manage even a symptomatic CM-I without surgery if
the symptoms are mild. For these reasons, we have not
emphasized patient symptoms in this analysis.

Any attempt to consider the natural history of CM-I
will need to account for selection bias in the decision to
pursue surgical treatment. The group of patients presented
here was selected for nonsurgical management. Therefore,
this analysis should only apply to the group of patients with
CM-I that is managed nonsurgically. Any conclusions that
may be derived from these asymptomatic or minimally
symptomatic patients should not be applied to symptom-
atic patients that are ordinarily considered good surgical
candidates. It is possible that the natural history is worse
for more symptomatic patients, for whom surgery is more
frequently offered. At our center, we do not follow a strict
protocol to determine if a CM-I will be treated surgically.
In general, we have offered surgery for patients with spi-
nal syrinx, patients with neurological deficits, and patients
with symptoms that are concordant with typical CM-I
symptoms and that have substantially interfered with the
patient’s quality of life. In some cases, patients were ini-
tially recommended for nonsurgical management and then
later underwent Chiari decompression despite a lack of any
new symptoms or radiological findings. In these cases, the
decision to offer surgery was made because symptoms had
persisted despite conservative management.

Our criteria for recommending surgical treatment of
CM-I must be considered in any analysis of those who
were treated without surgery at our institution. Based on
the results of this series, there is no basis for making any
assumptions about the natural history of the lesions in pa-
tients who did meet our usual surgical criteria and were
treated surgically. Given the widespread acceptance of
Chiari decompression in patients with either neurologi-
cal deficits or severe symptoms, it will be difficult to per-
form a natural history analysis of that group of patients
with CM-I. Nevertheless, we hope our analysis will help
to clarify the natural history of CM-I for the subgroup of
patients with CM-I that are considered to be asymptom-
atic or minimally symptomatic and without neurological
deficits. The mean duration of clinical follow-up by any
physician was only 6.5 years in this series. This follow-up
interval may be insufficient to capture all cases of clinical
or radiographic deterioration that could be seen over lon-
ger follow-up intervals. Further study over longer time in-
tervals will be necessary to clarify the natural history of
CM-I over the lifetime of individuals with this diagnosis.

The mean tonsillar descent is known to decrease with
advancing age in the normal population.25 This should be
considered in any interpretation of the natural history of
CM-I over long periods of time. Furthermore, it should
be noted that for this analysis, measurements of tonsillar
herniation were made without taking into account small
changes that may be seen in the degree of tonsillar descent
during the cardiac cycle.27,30 We believe that the changes in
tonsillar descent seen during the cardiac cycle (< 1 mm)
would not substantially alter our results or conclusions.
Conclusions
The natural history of CM-Is for those patients selected for conservative management is generally benign. Symptoms and MR imaging findings are stable over time in most cases, although spontaneous improvement and worsening of both the CM-I and the spinal syrinx do occur.

Disclosure
Dr. Muraszko is a consultant for Stem Cells, Inc. The remaining authors report no other conflicts 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: Maher, Muraszko, Garton, Strahle. Acquisition of data: Strahle, Kapurch, Maher, Muraszko, Garton. Analysis and interpretation of data: all authors. Drafting the article: Maher, Strahle. 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: Maher. Study supervision: Maher, Muraszko, Garton.

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