Pediatric and adult Chiari malformation Type I surgical series 1965–2013: a review of demographics, operative treatment, and outcomes

Aska Arnautovic, BSc,[1] Bruno Splavski, MD, PhD,[2] Frederick A. Boop, MD,[3,4] and Kenan I. Arnautovic, MD, PhD[3,4]

[1]George Washington University School of Medicine, Washington, DC; [2]Department of Neurosurgery, Osijek University School of Medicine, Osijek, Croatia; [3]Semmes-Murphey Clinic; and [4]Department of Neurosurgery, University of Tennessee, Memphis, Tennessee

OBJECT Chiari malformation Type I (CM-I) is a hindbrain disorder associated with elongation of the cerebellar tonsils, which descend below the foramen magnum into the spinal canal. It occurs in children and adults. Clinical symptoms mainly develop from alterations in CSF flow at the foramen magnum and the common subsequent development of syringomyelia.

METHODS The authors reviewed English-language reports of pediatric, adult, and combined (adult and pediatric) surgical series of patients with CM-I published from 1965 through August 31, 2013, to investigate the following: 1) geographical distribution of reports; 2) demographics of patients; 3) follow-up lengths; 4) study durations; 5) spectrum and frequency of surgical techniques; 6) outcomes for neurological status, syrinx, and headache; 7) frequency and scope of complications; 8) mortality rates; and 9) differences between pediatric and adult populations. Research and inclusion criteria were defined, and all series that contained at least 4 cases and all publications with sufficient data for analysis were included.

RESULTS The authors identified 145 operative series of patients with CM-I, primarily from the United States and Europe, and divided patient ages into 1 of 3 categories: adult (> 18 years of age; 27% of the cases), pediatric (≤ 18 years of age; 30%), or unknown (43%). Most series (76%) were published in the previous 21 years. The median number of patients in the series was 31. The mean duration of the studies was 10 years, and the mean follow-up time was 43 months. The peak ages of presentation in the pediatric studies were 8 years, followed by 9 years, and in the adult series, 41 years, followed by 46 years. The incidence of syringomyelia was 65%. Most of the studies (99%) reported the use of posterior fossa/foramen magnum decompression. In 92%, the dura was opened, and in 65% of these cases, the arachnoid was opened and dissected; tonsillar resection was performed in 27% of these patients. Postoperatively, syringomyelia improved or resolved in 78% of the patients. Most series (80%) reported postoperative neurological outcomes as follows: 75% improved, 17% showed no change, and 9% experienced worsening. Postoperative headaches improved or resolved in 81% of the patients, with a statistical difference in favor of the pediatric series. Postoperative complications were reported for 41% of the series, most commonly with CSF leak, pseudomeningocele, aseptic meningitis, wound infection, meningitis, and neurological deficit, with a mean complication rate of 4.5%. Complications were reported for 37% of pediatric, 20% of adult, and 43% of combined series. Mortality was reported for 11% of the series. No difference in mortality rates was seen between the pediatric and adult series.

CONCLUSIONS Before undergoing surgical treatment for CM-I, symptomatic patients and their families should be given clear information about the success of treatment and potential complications. Furthermore, surgeons may benefit from comparing published data with their own. In the future, operative CM-I reports should provide all details of each case for the purpose of comparison.

http://thejns.org/doi/abs/10.3171/2014.10.PEDS14295

KEY WORDS Chiari malformation Type I; pediatric; adult; series; demographics; treatment

Chiari malformation Type I (CM-I) is a cranio-cervical junction disorder that is associated with deformity and elongation of the cerebellar tonsils and is specifically characterized by tonsils' descent of more than 5 mm below the foramen magnum into the spinal canal. It occurs in children and adults and was first described by an Austrian pathologist, Hans Chiari, in 1891. The essential element in the pathophysiology of clinical symptoms is a change in the flow of CSF at the level of the foramen magnum, which is frequently associated with the subsequent development of syringomyelia.[10]

The incidence of tonsillar ectopia and CM-I has been reported to be 0.5%–3.5% in general, 0.56%–0.77% on MRI studies, and 0.62% in anatomical dissection studies,[20,98,99] which means that it is quite common in the general population. There seems to be a slight female predominance of cases (1.3:1), and the association with syringomyelia has been reported to range widely, from 30% to 70%.[90,127] Suboccipital craniectomy and foramen magnum decompression, with or without dural opening and with or without arachnoid opening or dissection, have been used most commonly for surgical treatment but without a clear idea of the frequency of use of either procedure in the reported series. Furthermore, it has not been noted whether a difference exists in the operative techniques used for pediatric and adult patients. Other factors that are also unclear are as follows: the geographical distribution of reports in the literature; the demographics of patients, including sex and age; the length of follow-up; the duration of study periods; the spectrum and frequency of surgical techniques used; neurological outcomes; improvements in outcomes for syrinx and headache; the frequency and scope of complications; mortality rates; and any differences between pediatric and adult populations with this malformation.

For this study, we reviewed the reports of pediatric, adult, and combined (adult and pediatric) surgical series of patients with CM-I published from 1965 through August 31, 2013, to delineate the timeline of the reported series and the number of reported patients and to investigate the factors listed above.

Methods

For this study, we searched PubMed/Medline for English-language reports of CM-I series (adult and pediatric) through August 31, 2013, using the key words "Chiari I malformation," "adult," "pediatric," and "series." In addition, we used references from these publications with the key term "Chiari I malformation" to locate series that did not have adequate key words to locate such reports in PubMed/Medline.

The research and inclusion criteria were defined before the study. We included series that contained at least 4 cases of CM-I and all publications with sufficient data for meaningful analysis. Certain series were published by the same authors at different times. To avoid an overlap of cases in the analysis, we included only the most recent publications of these authors if they were reports of consecutive series. If the same author(s) published different series, these were also included in the analysis. Two authors were involved in the study selection and 2 others in data extraction, with conference to determine consensus for discrepancies.

We excluded series of patients with achondroplasia, craniosynostosis, or combined Chiari Type I and II malformations that could not be mutually differentiated. We also excluded series in which patients with CM-I and syringomyelia were combined with patients whose syringomyelia was of a different origin and who therefore could not be mutually distinguished. In addition, we excluded patients with syringomyelia without CM-I, patients with acquired CM-I (i.e., after lumbar drainage), and series of patients with other congenital craniocervical malformations dominating, in addition to CM-I, with unclear mutual differentiation.

We analyzed the published CM-I series according to the year of publication and the country and continent of origin, and we categorized the cohort of patients into 1 of 3 groups: adult (> 18 years of age), pediatric (≤ 18 years of age), and combined/unknown. The lengths of follow-up were analyzed, including the medians and ranges. All the series were also analyzed according to age and sex distributions, and we determined the ranges, means, and standard deviations of these data. All the data were tabulated.

Operative treatment techniques were analyzed for the adult, pediatric, and combined patient cohorts, including suboccipital/foramen magnum decompression, dural opening, arachnoid opening, tonsillar resection, and CSF shunting. Among the adult, pediatric, combined, and total patient populations, postoperative improvement or resolution of suboccipital headache was analyzed when data were provided. Migraine headaches, neck pain, and other pain categories were not included. Postoperative neurological outcomes were most commonly listed as improvement/resolution, no change, or worsening. These outcomes in the adult, pediatric, combined, and total patient cohorts were examined separately. Postoperative syringomyelia outcomes were most commonly listed as improvement/resolution or no change. These outcomes in adult, pediatric, combined, and total patient cohorts were analyzed separately. Series that provided details of complications were analyzed for the adult, pediatric, and combined patient cohorts. Ranges and means of these data were noted, and series mortality rates were analyzed when they were provided in the report.

The frequency in differences of the investigated variables between the adult and pediatric series was determined with a chi-square or Fisher exact test. A 2-sided p value of < 0.05 was considered to be statistically significant. The Statistica for Windows 2005 program package version 7.1 (StatSoft, Inc.) was used for statistical analysis of the data.

Results

General Information

In the English-language literature published over a span of 48 years (1965–2013), we identified a total of 145 series of patients with CM-I, which included adult, pediatric, and combined adult and pediatric series (referred to here as combined series), in addition to series in which patient ages were not provided.[1–9,11–19,21–63,65–67,70,71,73–97,100,102–111,113–130,132–135,137–151,154–159] In this same span of published articles, we identified 40 series (27%) of adult patients[4,8,13,14,16–19,21,]

22,29,36,45,48,49,57,58,62,71,78,79,82,84,85,88,95,105,113,118–120,122–125,128,137,146, [149,151] and 39 series (27%) of pediatric patients[1–3,15,24,28,30–33,35, 41,42,50,51,54,56,60,61,65,77,80,81,86,89,92,93,97,104,108,109,116,133,134,143,145,150, 154,155] (Fig. 1). Combined series, which included both pediatric and adult patients, constituted 61 reports (42%).[5–7,9,11, 12,25–27,34,37,38,40,43,44,46,47,52,53,55,63,66,67,70,73,74,76,83,87,90,91,94,96,100,102, 103,106,110,111,114,115,117,121,126,127,129,130,132,135,138–142,144,147,148,156–159] We also identified 5 series (3%) in which the patients' ages could not be identified.[23,37,39,59,107] The median number of patients in each series was 31 (interquartile range [IQR] 15–68) and ranged from 4 to 585.

The durations of the studies ranged from 1 to 38 years, with a mean of 10 years (IQR 4–12 years). In the studies with pediatric patients, the mean study duration was 10 years (range 1–23 years), whereas in the studies with adult patients, the mean duration was 8 years (range 2–30 years). In the combined, pediatric, and adult series, the study durations had a mean of 11 years (range 1–38 years). The mean follow-up time of patients in all studies was 43 months (range 2–168 months). These data are shown in Table 1.

The largest number of studies was published over the past 21 years, from 1993 to 2013 (111 [77%] of 145 studies) (Fig. 2).

Geographical Distribution

The great majority of CM-I series were published in the United States,[1–3,5,6,8,9,15,16,18,19,22,24,25,27,30,31,33,35,38,41–43,46,50–54,56, 59,62,65,71,73,74,76,78,80,86,88–90,92,93,97,100,105–110,115,116,119,122,124,127,132, 133,143,150,151,154,155,157] followed by Great Britain,[7,39,40,70,91,94,96, 102,103,117,126,129,134,138] Italy,[28,36,57,60,114,118,120,137,141,145,149] France,[14,29, 47,61,67,104,111,135] and Japan.[55,75,79,113,128,130,139,142] There were 5 publications each from Spain,[58,121,146–148] Germany,[4,13,32,82,83] and Turkey[48,49,84,85,156] and 4 publications from Brazil.[12,26, 125,140] Three studies each were from Canada[44,77,95] and China.[66,158,159] Poland,[21,87] Belgium,[34,123] and India[11,63] each

had 2 publications. The remaining countries published a report on 1 series each[17,23,37,45,81,144] (Figs. 3 and 4).

Demographics

The total number of patients in all the published series was 8605: 2351 adult (27%), 2583 pediatric (30%), and 3671 unknown (43%)[1–9,11–19,21–63,65–67,70,71,73–97,100,102–111,113– 130,132–135,137–151,154–159] (Fig. 5).

In the adult-only series,[4,8,13,14,16–19,21,22,29,36,45,48,49,57,58,62,71,78, 79,82,84,85,88,95,105,113,118–120,122–125,128,137,146,149,151] there were 1608 patients: 543 men (34%), 913 women (57%), and 152 (9%) of unknown sex.

In the pediatric-only series, there were 2302 patients: 578 boys (25%), 635 girls (28%), and 1089 (47%) of unknown sex.[1–3,15,24,28,30–33,35,41,42,50,51,54,56,60,61,65,77,80,81,86,89,92,93,97,104, 108,109,116,133,134,143,145,150,154,155]

In the published series, cases in females predominated over those in males (39% vs 29%, respectively); there were 57% women versus 34% men in the adult series and 28% girls versus 25% boys in the pediatric series (p < 0.001, Fisher exact test) (Fig. 6).

The median age of adult patients was 40.5 years (IQR 37–45.3 years), of pediatric patients, 8 years (IQR 6–10.5 years), and in the combined studies, 35 years (IQR 27.3– 40 years) (Table 2). There were 2 peak ages of presentation in both the pediatric and the adult series. The most common (peak) age presentation in the pediatric series was 8 years, followed by 9 years (Fig. 7 upper). The most common (peak) age presentation in the adult series was 41 years, followed by 46 years (Fig. 7 lower).

Incidence of Syringomyelia

Of a total of 6347 patients in all operative series for whom an association with a syrinx was reported, 4144 (65%) harbored a spinal cord syrinx. The range of syringomyelia in this series ranged from 12% to 100%. The incidence of syringomyelia ranged from 20% to 100% in the adult series, 12% to 100% in the pediatric series, and 32% to 100% in the combined series[1–4,6–9,12–19,21–25,27–37,40–42,45–58,60–63,65–67,70,71, 73–77,79,81–87,89,92–97,100,102–106,108–111,113–115,117–130,132–135,137,139,140,142–151, 154,155,157–159] (Table 3).

In the adult series that reported syrinx association, 1090 (69%) of 1578 patients had a syrinx.[4,8,13,14,16–19,21,22,29,36,45,48,49, 57,62,71,79,82,84,85,95,105,113,118–120,122–125,127,128,137,149,151] In the pediatric series that reported an association of syringomyelia, of a total of 1678 patients, 673 (40%) harbored a syrinx.[1–3,15, 24,28,30–33,35,41,42,50,51,54,56,60,61,65,77,81,86,89,92,93,97,100,104,108,109,133,134,145, 150,154,155] In the combined series that reported this association, of a total of 3091 patients, 2411 (78%) had a syrinx.[6,7,9, 12,23,25,27,34,37,40,46,47,52,53,55,58,63,66,67,70,74–76,87,94,96,100,102,103,106,110,111, 114,115,117,121,126,129,130,132,135,138–140,142–144,147,148,157–159] Significantly more common was the incidence of syringomyelia in the adult series (p < 0.001, Fisher exact test) (Fig. 8).

Operative Techniques

Of a total of 140 adult, pediatric, and combined studies (5 of 145 series were unknown and, thus, not included),[1–9, 11–19,21,22,24–36,38,40–58,60–63,65–67,70,71,73–97,100,102–106,108–111,113–130,132– 135,137–151,154–159] only 4 did not describe the operative techniques used. Of the 136 series that reported operative
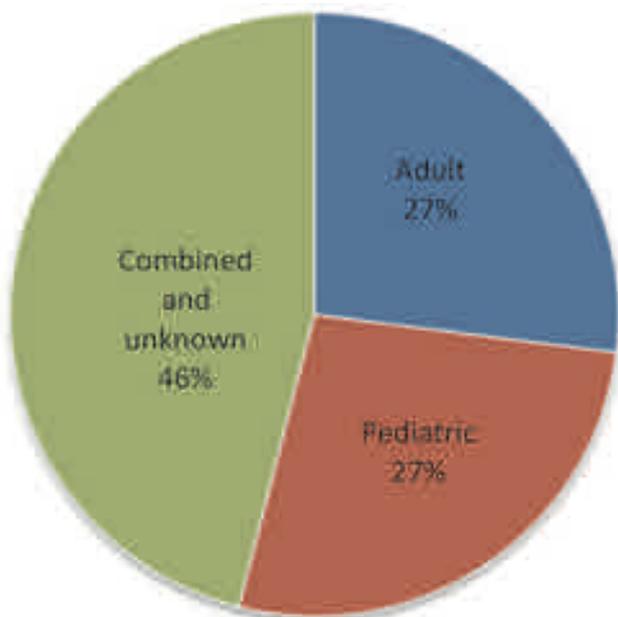

FIG. 1. Age distribution of patients in the CM-I operative series.

TABLE 1. Follow-up times

Study Type	Follow-Up Time (mos)			
	Mean (SD)	Min/Max	Median	IQR
Adult series	46 (31.5)	3/151	28	14–52
Pediatric series	37 (26.8)	2/120	39	16–49.5
Combined series	39 (36.3)	6/168	40	17.5–60.5
Total	43 (35.2)	2/168	39	17–51

techniques, 134 (99%) reported the use of posterior fossa/foramen magnum decompression. In 123 (92%), the dura was opened. In 81 (66%) of these studies, the arachnoid was opened and dissected, and in 22 (27%) of these 81, tonsillar resection was performed, whereas some sort of syrinx shunting was carried out in 25 patients.

Of 39 pediatric series,[1–3,15,24,28,30–33,35,41,42,50,51,54,56,60,61,65,77,80,81,86,89,92,93,97,104,108,109,116,133,134,143,145,150,154,155] 38 (97%) delineated the operative techniques used. Of these, 37 studies (97%) described the use of posterior fossa decompression. One study each (2.7%) reported the use of endoscopic decompression, tonsillectomy without craniectomy, and
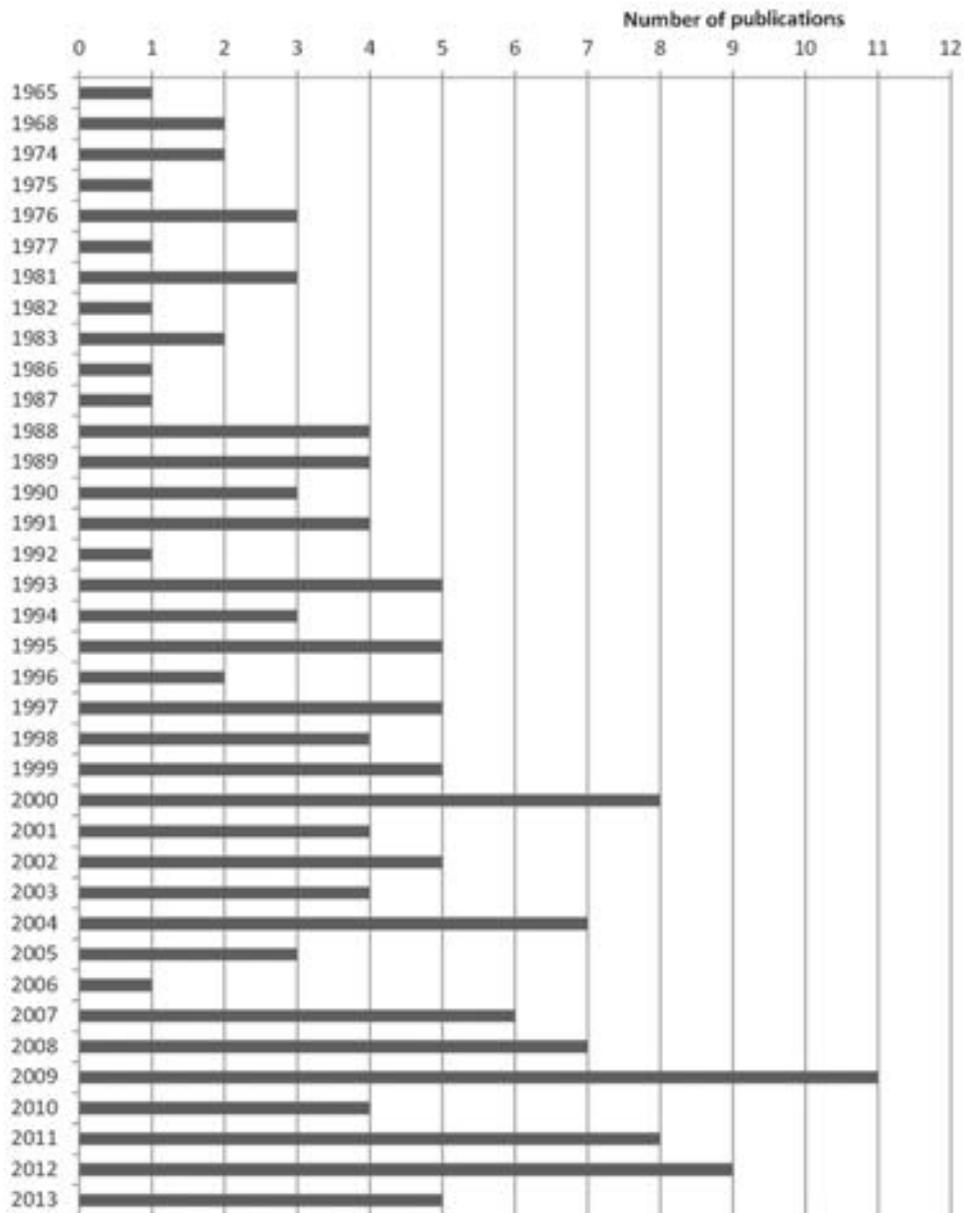

FIG. 2. Number of CM-I operative studies according to the year of publication.
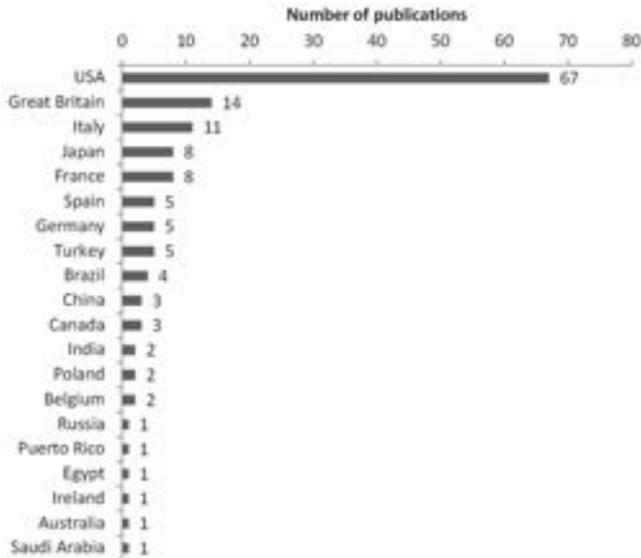
FIG. 3. Number of operative CM-I studies according to the country of publication.

FIG. 5. Age distribution of patients with CM-I.

cranioplasty after decompression. Of 37 studies involving posterior fossa decompression, 30 (81%) reported dural opening, and of them, 14 (47%) added arachnoid opening and dissection. In 4 series, some sort of shunting procedure was added. Six (43%) of the 14 studies that described arachnoid opening and dissection added tonsillar resection as well.

Of 40 adult series,[4,8,13,14,16–19,21,22,29,36,45,48,49,57,58,62,71,78,79,82,84,85,88,95,105,113,118–120,122–125,128,137,146,149,151] 37 (93%) reported the
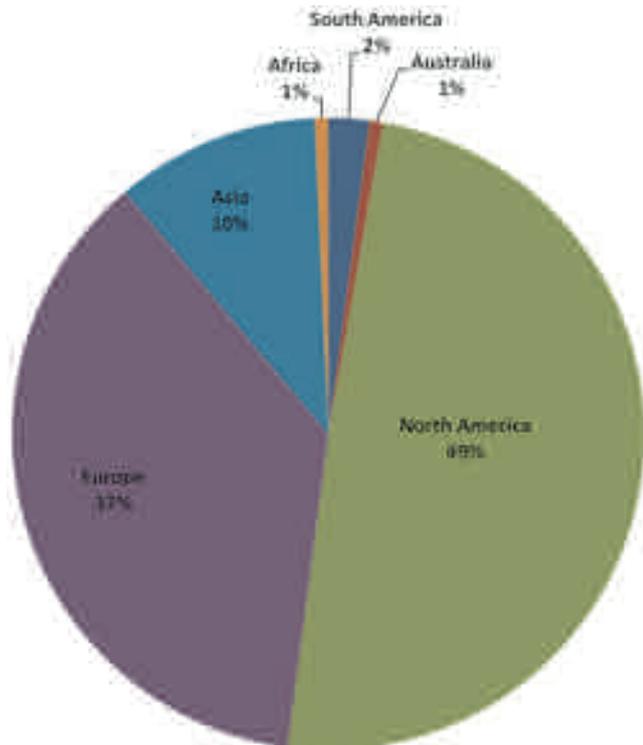

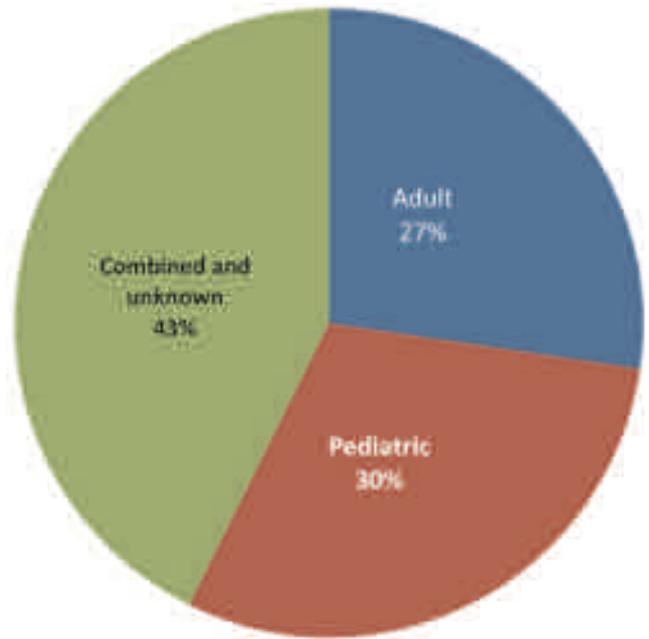**FIG. 4.** Distribution of CM-I studies according to the continent of publication.

operative techniques used. All of them described the use of posterior fossa decompression. All but 1 reported dural opening (97.3%), and 3 studies (8%) reported only dural relaxation incisions. Of all the series that reported the use of posterior cranial fossa/foramen magnum decompression, 26 (70%) added arachnoid opening and dissection, and of them, 6 (23%) added tonsillar resection. In 2 series, syrinx shunting was also reported.

All 61 combined series[5–7,9,11,12,25–27,34,37,38,40,43,44,46,47,52,53,55,63,66,67,70,73,74,76,83,87,90,91,94,96,100,102,103,106,110,111,114,115,117,121,126,127,129,130,132,135,138–142,144,147,148,156–159] reported the operative techniques used. All but 1 of these (98%) reported the use of suboccipital craniectomy/posterior fossa decompression/foramen magnum decompression. Only 1 reported syrinx shunting. Of all the series using suboccipital craniectomy/foramen magnum decompression, the dura was opened in 57 (95%) of the studies, and of these 57, 41 (72%) reported arachnoid opening/dissection. Of the 41 studies that reported arachnoid opening/dissection, 10 (24%) added tonsillar resection. Shunting of the syrinx was performed in 25 patients (Table 4).

Postoperative Syringomyelia Outcomes

Postoperative improvement of syringomyelia was reported for 1108 (78%) of 1423 patients.[1–4,6–9,12–17,21–25,27–37,40–42,45–58,60–63,66,67,70,73–75,79–87,89,92–94,96,100,102–106,108–111,113–115,117,119,120,122–130,132–135,137–145,147–150,156–159] It should be noted that of all the series for which an association of syringomyelia with CM-I was reported, only some postoperative syringomyelia outcomes were reported; hence, there was disproportion between reporting syringomyelia association and syringomyelia outcomes.

In the adult series, syrinx improvement was reported for 293 (78%) of 375 patients with a syrinx. The reported ranges for individual adult series were 38%–100% improvement/resolution, 11%–64% no change, and 2%–40% worsening.
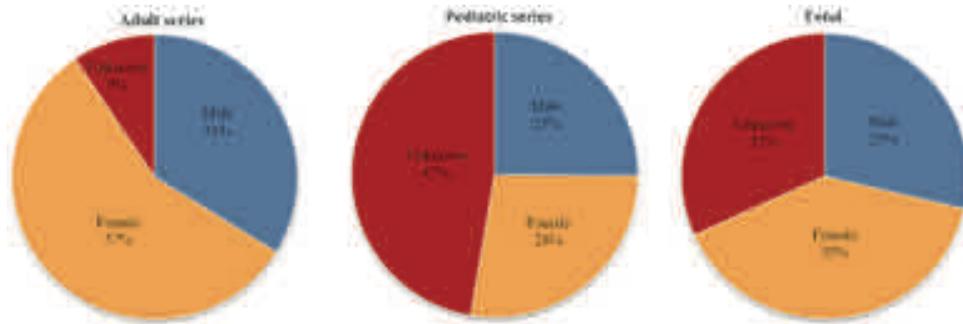
FIG. 6. Sex distribution of patients with CM-I.

In the pediatric series, improvement/resolution of the syrinx ranged from 50% to 100% of patients; 163 (79%) of 207 patients had syrinx improvement. Patients with no change ranged from 8% to 50%, and an increase in size (worsening) occurred in 5%–12%.

In the combined series, improvement/resolution of the syrinx was reported to range from 39% to 100%, with no change in syrinx size in 3%–42% and an increase in the syrinx size (worsening) in 3%–45%. In these series, syrinx improvement/resolution was reported for 1108 (78%) of 1423 patients. There was no difference in the syrinx improvement/resolution rates between the adult and pediatric series (p = 0.916, Fisher exact test) (Table 5, Fig. 9).

Postoperative Neurological Outcomes

The majority of series (116 series [80%]) reported postoperative neurological outcomes as improvement/resolution, no change, and worsening of neurological status. It should be noted that these evaluations were performed subjectively by the treating surgeons and reported by the publications' authors. Of 4206 patients, 3150 (75%) were reported to be improved, 697 (16.6%) had no change in neurological status, and 359 (8.5%) experienced worsening.[3–7,11,13,15,17–19,21–23,25–37,40–46,48,49,52,54–56,58,59,62,63,65–67,70,71,73–87,89–91,93–97,100,102,105,106,109–111,113–121,123–125,127–130,133,135,137–151,154–159]

Of 1126 patients in the adult series, improvement/resolution was noted in 823 (73%), no change in 225 (20%), and worsening in 78 (7%). The range of improvement/resolution in individual series was 22% to 100%, no change in status ranged from 4% to 65%, and worsening ranged from 5% to 43%.[4,13,17–19,21,22,29,36,45,48,49,58,62,71,78,79,82,84,85,95,105,113,118–120,123–125,128,137,149,151]

The pediatric series reported improvement/resolution of status in individual series ranging from 55% to 100%, no change in 5%–30%, and worsening in 5%–18%. Improvement/resolution was reported for 669 (84%) of 796 patients, no change in 116 (15%), and worsening in 11 (1%).[3,15,28,30–33,35,41 42,54–56,77,80,81,86,89,93,97,109,133,143,145,150,154,155]

The combined pediatric and adult series reported improvement/resolution to range in individual series from 27% to 100%, no change to range from 4% to 64%, and worsening to range from in 2% to 45%. In these series, of a total of 2284 patients, improvement/resolution was noted in 1658 (72%), no change in 356 (16%), and worsening in 270 (12%).[5–7,11,23,25–27,34,37,40,43,44,46,52,55,58,63,66,67,70,74–76,87,90,91,94,96,100,102,106,110,111,114,115,117,121,127,129,130,135,138–140,142–144,147,148,157–159]

There was significantly greater worsening of neurological outcomes in the adult series than in the pediatric series (p < 0.001, chi-square test) (Table 6, Fig. 10).

Postoperative Headache Outcomes

Postoperative headache outcomes categorized by improvement/resolution were reported for 23 series (16%), in 350 (81%) of a total 433 patients.[1,2,11,19,32,37,43,55,60,62,70,73,80,86,89,93,108,110,118,127,138,142,150] Of a total of 179 pediatric patients, improvement/resolution of headache was noted in 157 (88%). Improvement/resolution in the individual series ranged from 66% to 100%. Of 128 adult patients, improvement/resolution was noted in 93 (73%). In individual pediatric series, improvement/resolution ranged from 37% to 100%, whereas in the combined adult and pediatric series, of 126 patients, improvement/resolution was noted in 100 (79%). In the individual series, it ranged from 12% to 92% (Fig. 11). There was a significant difference in improvement/resolution of headache in favor of the pediatric series over that of the adult series (p = 0.001, Fisher exact test).

Postoperative Complications

Postoperative complications were noted in 60 (41%) of 145 series.[2,4,6,11,12,15,18,19,24,25,29,32,35,38,41,42,44,45,48,49,52–54,56,60,66,70,75,76,82,83,85–88,90,92,93,96,97,104–106,109,115–118,120,126,133,135,140,143,145,146,148,150,155,156,159] In these series, the median complication rate was 4.5% (IQR 3.3%–7%). The complications most commonly reported were CSF leak (range 1% to 24% [median 5%; IQR 2%–10.5%]), pseudomeningocele (range 1%–47%

TABLE 2. Patient ages in CM-I studies

Study Type	Patient Age (yrs)			
	Mean (SD)	Min/Max	Median	IQR
Adult series	40.9 (5.9)	18/72	40.5	37–45.3
Pediatric series	7.7 (3)	0.25/18	8	6–10.5
Combined series	33 (12)	0.3/70	35	27.3–40
Total	27 (8)	0.25/72	36	27–40

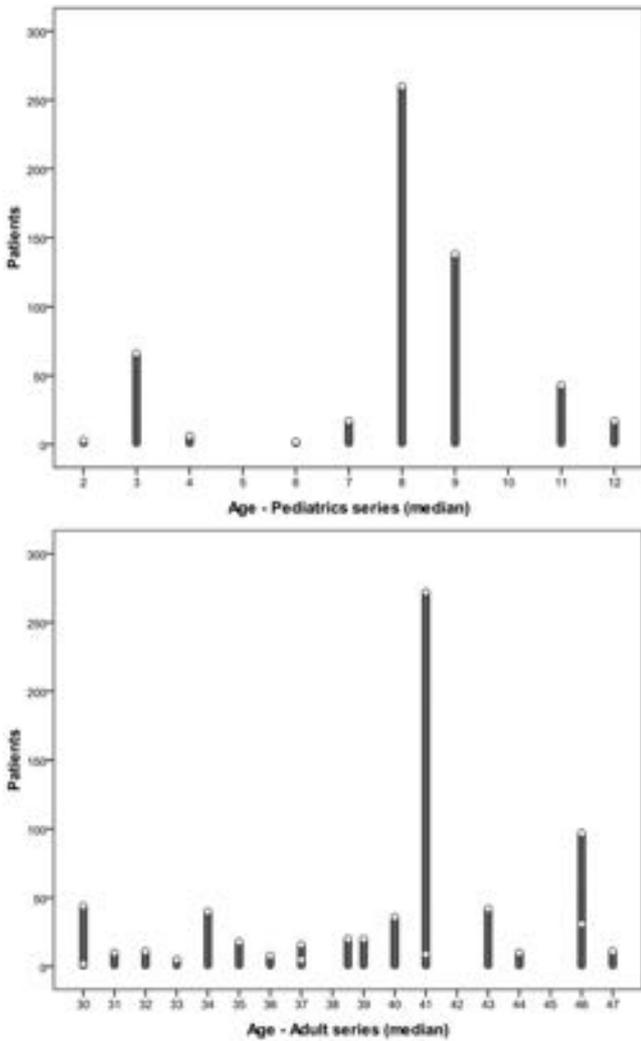FIG. 8. Incidence of syringomyelia in patients in the CM-I operative series.

FIG. 7. **Upper:** Scatter plot graph of the presentation ages of patients with CM-I in the pediatric series. **Lower:** Scatter plot graph of the presentation ages of patients in the adult series.

[median 12%; IQR 3%–20%]), aseptic meningitis (range 1%–40% [median 4.5%; IQR 3%–8%]), wound infection (range 1%–11% [median 3%; IQR 2%–5.5%]), meningitis (range 1%–6% [median 3.5%; IQR 2%–6.3%]), neurological deficit (range 1%–8% [median 4%; IQR 4%–6%]), epidural/subdural hematoma of the posterior cranial fossa (range 1%–2%), hydrocephalus and respiratory dysfunction (median 2%; IQR 1%–2% each), and postoperative headaches (in 9% of cases in 1 series). Of 60 series reporting complications, 22 (37%) were pediatric, 12 (20%) were adult, and 26 (43%) were combined (Fig. 12).

In the pediatric series, complications were reported in 22 of 39 studies (56%), with a median of 3.5% (IQR 2.3%–

14.3%). The complications most commonly reported were pseudomeningocele (range 5%–47% [median 15%; IQR 4.5%–18%]), aseptic meningitis (range 1%–60% [median 6%; IQR 3%–21%]), CSF leak (range 0.2%–21% [median 4%; IQR 3%–10.5%]), wound infection (range 1%–9% [median 3%; IQR 1%–4%]), meningitis (range 2%–4% [median 2%; IQR 2%–3%]), neurological deficit (range 1%–4% [median 2%; IQR 1%–3%]), epidural/subdural hematoma of the posterior cranial fossa (range 1%–2%), and hydrocephalus and respiratory insufficiency in 2% each. In 9% of the cases in 1 series, postoperative headaches were reported as a complication.

Postoperative complications in 12 of the adult studies were noted, with a median of 3% (IQR, 2.5%–5.5%). The most common were pseudomeningocele (range 1%–40% [median 2.5%; IQR 1.8%–12%]), aseptic meningitis (range 2%–4% [median 3%; IQR 2.5%–3.5%]), CSF leak (range 2%–11% [median 3%; IQR 1%–5%]), and meningitis (range 1%–4% [median 3%; IQR 2%–3.5%]) (Fig. 13).

The combined adult/pediatric series reported complications in 26 (60%) of 43 studies, with a median of 3% (IQR 1.3%–3.8%). The most common complications were CSF leakage (range 1%–24% [median 6%; IQR 5%–13%]), pseudomeningocele (range 1%–23% [median 12%; IQR 5%–18%]), aseptic meningitis (range 1%–33% [median 7%; IQR 4%–11%]), wound infection (range 2%–11% [median 3%; IQR 2%–7%]), meningitis (range 3%–9% [median 7%; IQR 5%–8%]), neurological deficit (range 1%–8% [median 4%; IQR 4%–5%]), epidural hematoma of the posterior cranial fossa (1%), trigeminal neuralgia (3%), intraventricular bleeding (3%), respiratory dysfunction (1%), and embolization (2%).

TABLE 3. Incidence of syringomyelia in CM-I operative series

Study Type	No. (%)	Range (%)
Adult series	1090/1578 (69)	20–100
Pediatric series	673/1678 (40)	12–100
Combined series	4144/6347 (65)	32–100

TABLE 4. Distribution of operative techniques in CM-I

Technique Used	Adult Series (n = 40) (no./total no. [%])	Pediatric Series (n = 39) (no./total no. [%])	Combined Series (n = 61) (no./total no. [%])
Operative technique	37/40 (93)	38/39 (97)	61/61 (100)
Posterior fossa/foramen magnum decompression	37/37 (100)	37/38 (97)	60/61 (98)
Dural opening/duraplasty	36/37 (97)	30/37 (81)	57/60 (95)
Arachnoid opening/dissection	26/37 (70)	14/30 (47)	41/57 (72)
Tonsillar resection	6/26 (23)	6/14 (43)	10/41 (24)
Shunt	2/26 (8)	4/30 (13)	19/60 (32)

Mortality

Sixteen studies (11%) reported mortality-related data.[2,8,11,17,23,26,32,58,70,90,95,96,102,117,140,159] Mortality was reported for 2 of 65 (3%) pediatric patients (years of study publication 1994 and 2010). Mortality was reported for 3 of 204 adult patients (2%) (years of study publication 1974–1977) and for 30 of 990 patients (3%) in the combined series (years of study publication 1968–2008). There was obviously no difference in the mortality rates between the adult and pediatric studies. The most commonly reported etiologies were pneumonia/respiratory failure (9 patients), infection/sepsis (7 patients), postoperative bleeding (3 patients), and sleep apnea (2 patients), whereas the rest of the etiologies were unknown or unclear.

Discussion

Chiari malformation Type I is a fairly common condition in the general population, ranging from 0.5% to 3.5% in general, from 0.56% to 0.77% in MRI, and 0.62% in anatomical brain-dissection studies.[20, 98, 99] Among patients with CM-I, associated syringomyelia is frequent and has been reported to range from 30% to 70%.[90, 98, 99, 127] Between 15% and 30% of adult patients with CM-I are reportedly asymptomatic.[20]

Suboccipital craniectomy and foramen magnum decompression, with or without dural opening and with or without arachnoid opening and dissection, have been used most commonly for operative treatment; however, there is no clear idea of the frequency of their use in the reported series or whether a difference exists between pediatric and adult patients. Other unknowns are the precise geographic and continental distributions of reports of this malformation, sex and age demographics, lengths of follow-up, study durations, neurological outcomes, syrinx and headache improvement outcomes, rate and scope of complications, mortality, and whether differences exist between pediatric and adult populations with this malformation.

We encounter fairly large numbers of adult and pediatric patients with CM-I in our practice. Symptomatic patients and their families regularly ask precise questions regarding the expected success of their prospective operative treatment, the chances for improvement of their neurological deficits and syringomyelia, headaches, and the risks of potential complications. Furthermore, surgeons may benefit from comparing to their own patients the published demographics, operation types, outcomes, and complications in these patients. Our residents and members of other medical specialties who participate in the care of patients with CM-I frequently ask questions about various aspects of demographics, treatments, and outcomes. Finally, while preparing publications and presentations about various aspects of CM-I, many precise data are not available. These multiple reasons prompted this extensive review to provide all needed data in a single publication. To our knowledge, there has not been conducted thus far such an extensive and thorough review of operative CM-I series in the literature.

We identified 145 operative series of CM-I over the past 48 years (1965–2013).[1–9,11–19,21–63,65–67,70,71,73–97,100,102–111,113–130,132–135,137–151,154–159] Most of these publications (76%) were published in the past 21 years, with a clear increase in the frequency of publications in recent years. Thus, the results presented herein are fairly contemporary and present, in a broad sense, the current state of the art of operative treatment (Fig. 2). This increased frequency of reporting clearly coincides with the development and advancement of MRI techniques. The use of this diagnostic modality has undoubtedly contributed to the earlier and more accurate diagnosis of this condition and its severity, easier and more precise clinical follow-up of patients who are treated operatively and nonoperatively, improvements in the understanding of CM-I pathophysiology, refinement of operative treatments and outcomes, and an understanding of postoperative complications and the possibilities for preventing them.

Study Limitations

This review article analyzes the reported case series of patients with CM-I containing mostly Class III and, in rare

TABLE 5. Postoperative syringomyelia outcomes in patients with CM-I

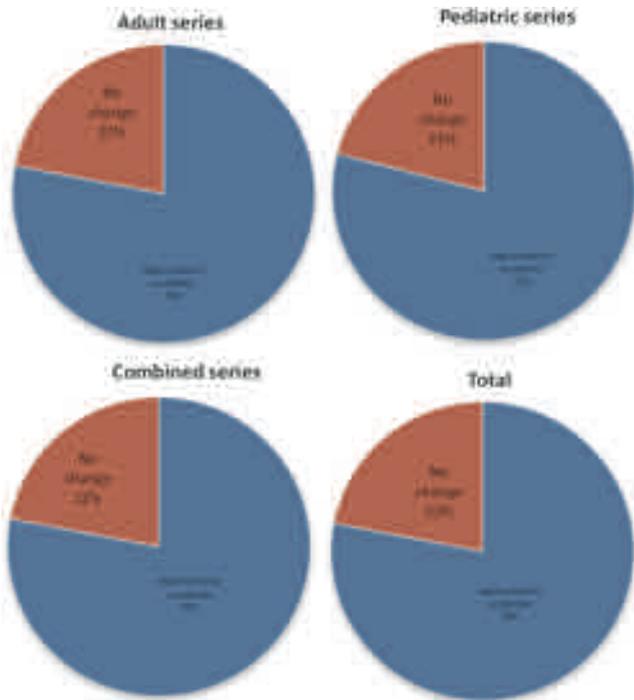| Study Type	Postop Syrinx Outcomes in % (range)		
	Improvement/Resolution	No Change	Worsening
Adult series	38–100	11–64	2–40
Pediatric series	50–100	8–50	5–12
Combined series	39–100	3–42	3–45

FIG. 9. Distribution of patients with CM-I according to syringomyelia outcome.

cases, Class II evidence. This analysis was based on operative CM-I series only and did not include nonoperative series. These series have been published at different times, in different scientific journals, and from different countries. Non–English language literature on the subject was not included. As such, our study bears a number of limitations pertaining to these facts. While considering the strength of the information presented in this review (mostly Class III), the above facts should be taken in consideration.

Geographics

Almost half of the publications (46%) dealing with CM-I operative treatment for CM-I were published from the United States, and the majority of the other half came from the European Union (33%). The remaining reports originated from the rest of the world. It was surprising that Australia and Africa (Egypt) had only 1 publication each (Figs. 3 and 4). It is quite improbable that there is an increased incidence of CM-I in North America and Europe compared with that in the rest of world, and the reason for the disparity of reports from these and other regions is probably multifactorial. Some of the likely factors are as follows: a higher socioeconomic status and subsequent higher level of health care in North America

and Europe, with an increased ratio of physicians, neurologists, and neurosurgeons to the population; a larger number of MRI facilities in North America and Europe; and a greater number of medical journals in North America and Europe available to publish reports. An increased incidence of patients with CM-I having a higher body mass index (BMI) and the influence of BMI on the formation of syringomyelia was elaborated recently.[10] Therefore, one may speculate that a higher BMI is more common in wealthier nations. Furthermore, there is the question of whether a higher average age of the population in certain regions plays a role in the higher incidence of CM-I. Racial and ethnic differences may also play a role. In addition, medicolegal ramifications may influence the frequency with which studies are ordered; thus, more symptomatic patients may be discovered.

Study Durations, Patient Numbers, and Lengths of Follow-Up

The median number of patients per series was 31 (range 4–585). The mean study duration was 10 years (IQR 4–12 years), but it was somewhat shorter for adult patients (8 years). The mean follow-up time of all the patients was 43 months (slightly more than 3.5 years), but it was longer in adult series (46 months) than in the pediatric and combined series (37 and 39 months, respectively).[1–9,11–19,21–63,65–67,70,71,73–97,100,102–111,113–130,132–135,137–151,154–159] The distribution of follow-up times was asymmetrical, with median follow-up time shorter that the mean, showing that most of the studies had shorter follow-up times.

These findings suggest that, on average, investigators needed a relatively long period of time for the study (10 years), a moderately long mean follow-up time (3.5 years), and a moderate number of patients (31 cases) to accrue enough data for an individual study publication. These findings also point to the fact that, after surgery, most patients (adult and pediatric) are released from care after a maximum of 3.5 years (even less in some series) and that, for most patients, a follow-up time longer than 3.5 years is not needed.

Age of Patients

The distribution of adult and pediatric series (Fig. 1) and the ages of the adult and pediatric patients (Fig. 5) show that the frequency of CM-I has been similar between these 2 groups, with pediatric patients having just slightly more instances of the malformation. Therefore, it seems that CM-I is not more common in any of the age groups. The mean and median ages in the pediatric patient population were both 8 years, whereas in the adult population, they were 41 years. The mean and median ages in the combined studies were 33 and 35 years, respectively. In all the studies, the overall mean and median ages were 27 and 36

TABLE 6. Postoperative neurological status outcomes in patients with CM-I

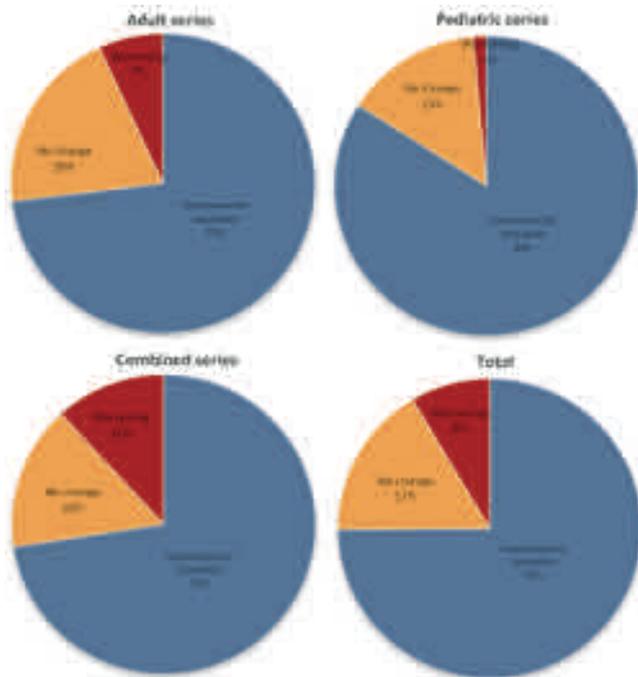| Study Type	Postop Neurological Outcomes in % (range)		
	Improvement/Resolution	No Change	Worsening
Adult series	22–100	4–65	5–43
Pediatric series	55–100	5–30	5–18
Combined series	27–100	4–64	2–45

FIG. 10. Postoperative distribution of neurological status outcomes according to CM-I series.

years, respectively (Table 2). It is interesting to note that 2 peak ages of presentation were delineated for each population (i.e., pediatric and adult). The peak ages of presentation in the pediatric studies were 8 years, followed by 9 years (Fig. 7 upper). The peak ages of presentation in the adult series were 41 years (Fig. 7 lower), and this figure was reported earlier.[64,90] However, the next peak age was 46 years, which is a new finding. These data may help phy-
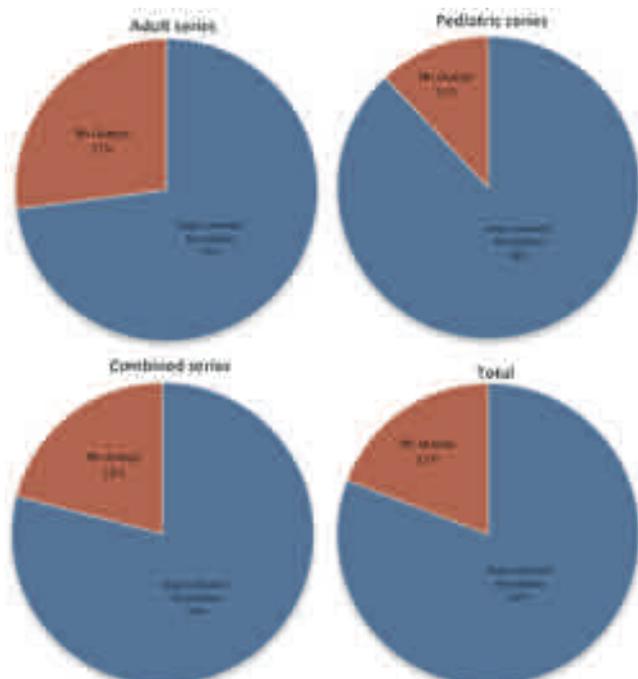

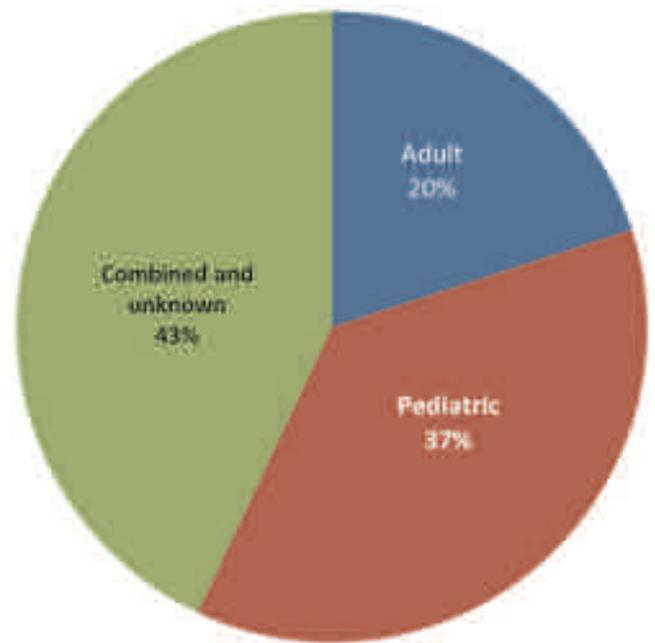FIG. 11. Postoperative headache outcomes according to CM-I series.

FIG. 12. Distribution of reporting complications in the CM-I series.

sicians to keep focus on the diagnostics in symptomatic patients of corresponding ages.

Sex of Patients

We noted a statistical difference between the sexes of patients in the studies, with females (39%) predominating (57% women [adult series] and 28% girls [pediatric series]). One reason for this predominance might be that women, particularly in the adult population, present more commonly with headache syndrome, which in turn may prompt more frequent MRI diagnostics. Meadows et al.[98] questioned whether the female predominance is the result of more frequent initial cerebellar ectopia or a faster progression of symptoms later in life. Our data indicate that both are probably true.

Incidence of Syringomyelia

Syringomyelia is a cavitary enlargement[101] or cyst formation[71] of the spinal cord. CM-I is the leading cause of syringomyelia, and its pathogenesis was initially established by 2 classic hypotheses of Gardner[59] and Williams.[152,153] Milhorat et al.[101] did experimental work on syringomyelia and the central spinal canal. More recently, Heiss et al.[71,72] and Oldfield et al.[112] proposed new insights into the pathophysiology of syringomyelia and its implications for the diagnosis and treatment of CM-I. Obviously, the creation and enlargement of a syrinx may contribute to the transformation of asymptomatic to symptomatic CM-I. In addition, gaining weight has been proposed as one of the contributing factors for de novo syrinx creation and patients becoming symptomatic.[10]

The association of syringomyelia with CM-I has been reported to range from 30% to 70%[64] and 37% to 75%.[27,79,98,99,110,117] This review also demonstrates the exact association of syringomyelia with CM-I in operative series. Ranges were 20%–100% in the adult series, 12%–100%
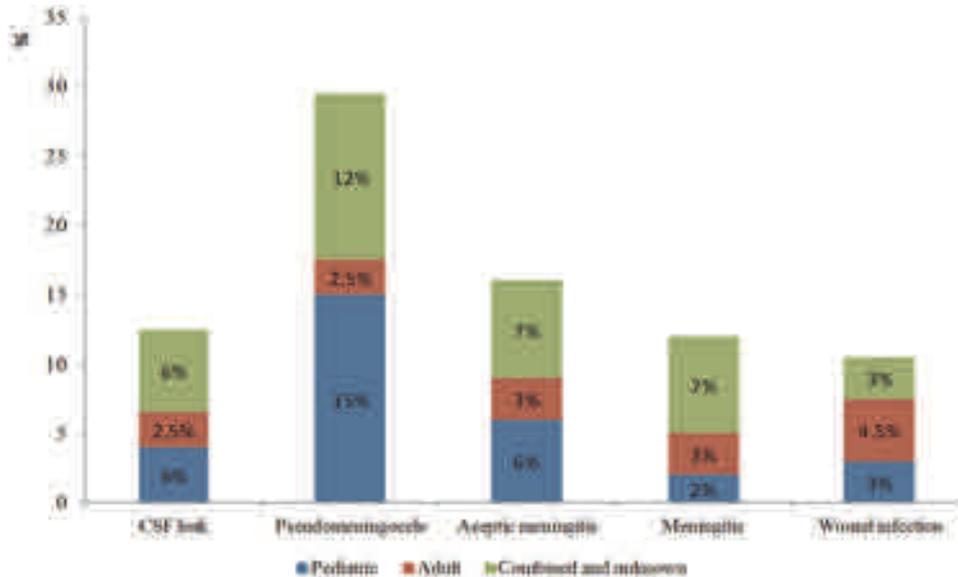
FIG. 13. Postoperative complications of CM-I; the distribution of the medians is stratified according to series.

in the pediatric series, and 32%–100% in the combined series. In absolute numbers, the rates of association of syringomyelia with CM-I were 69% in adult patients, 40% in pediatric patients, 78% in patients in the combined series, and 65% overall. In other words, two-thirds of all patients with CM-I treated operatively had syringomyelia (Table 3, Fig. 8), a statistically significant difference in favor of syrinx presence versus absence in patients with CM-I. This is not a surprise, because a syrinx gradually develops over years in a patient with CM-I. This frequency of syringomyelia in the series of patients with CM-I treated operatively confirms the notion that the presence of a syrinx plays one of the most important roles in symptomatology, the severity of presentation, and indications for surgery in all patient populations. Furthermore, the data on the mean and median ages of presentation (27 and 36 years, respectively) found in all the series and the fact that syringomyelia is more common in adult than in pediatric patients support the fact that symptomatic CM-I presents relatively late in life and needs time to develop. This fact is particularly interesting considering congenital and embryological theories of the development of this disorder (Table 2).

Operative Techniques

Several review articles surveyed the treatment of syringomyelia and CM-I in pediatric practices,[69,131] but we were not able to identify similar surveys of adult CM-I practices. These reports by Haroun et al.[69] and Schijman and Steinbok[131] acknowledged differences in the management of pediatric CM-I. One consensus among these reviews was that asymptomatic patients should be observed unless there is an associated syrinx, and that most patients with CM-I with scoliosis underwent decompression regardless of whether they had an associated syrinx. Suboccipital foramen magnum decompression was the standard surgical procedure used, with the majority of surgeons opening the dura and using a graft. In their reviews of pediatric CM-I publications, Durham and Fjeld-Olenec[42] and Hankinson

et al.[68] reported that posterior fossa decompression and dural opening with duraplasty are associated with a lower risk of reoperation but a greater risk of CSF-related complications. Sindou and Gimbert[136] compared their technique of dural opening with duraplasty and preservation of the arachnoid membrane with other reported techniques. A Japanese review examined the differences between orthopedic (posterior fossa decompression only) and neurosurgical (posterior fossa decompression with duraplasty) techniques of decompression. They reported that the neurosurgical approach was associated with better neurological and syrinx improvement outcomes.[130]

The main goal of surgery for CM-I was summarized by Batzdorf[18] as follows: resolving craniospinal pressure dissociation, restoring subarachnoid spaces and the cisterna magna in the posterior cranial fossa, eliminating and reducing the syrinx, relieving compression of the brainstem, and relieving or eliminating symptoms and signs of CM-I. In our analysis of published operative CM-I series, we noted that all the combined series, 97% of the pediatric series, and 85% of the adult series reported the operative technique used for treatment, and almost all of them described the use of posterior fossa/foramen magnum decompression. Furthermore, the dura was opened in 95%, 81%, and 97% of the cases, respectively. In addition, opening of the arachnoid membrane and intra-arachnoid dissection were reported in 72% and 70% of the combined and adult series, respectively, but only 47% of the pediatric series. The reason for the lower frequency of arachnoid membrane opening in pediatric studies probably lies in the attempt of some authors to decrease the incidence of CSF-related complications (pseudomeningocele, CSF leak, and aseptic and bacterial meningitis). However, considering all the studies (Table 4), the majority of patients were treated with arachnoid opening/dissection. We may speculate that there were 2 reasons for this treatment choice. One is that there may have been an arachnoid membrane velum closing the Magendi foramen, and most surgeons opted to directly in-

spect and open this membrane as needed. The other reason may be that according to their experience, a majority of the surgeons correlated overall good results of CM-I treatment with adding the arachnoid opening/dissection.

Tonsillar resection was relatively common in the pediatric series. Almost half of the pediatric studies (43%) in which the arachnoid membrane was opened also included this treatment, whereas only about one-quarter of adult and combined studies in which the membrane was opened (23% and 24%, respectively) added tonsillar resection. The reason for the more frequent tonsillar resection in pediatric patients is unclear.

Shunting was rarely performed, but it was used most commonly in the combined series (32%). One may speculate that some authors historically did not achieve the desired results with syrinx resolution after surgical decompression alone and therefore added shunting for their patients. Only a small proportion of procedures involved bone decompression only, possibly because of the relatively recent advent of that procedure.

Overall, posterior fossa/foramen magnum decompression, dural opening with duraplasty, and arachnoid opening with dissection were the most commonly used procedures in the adult and combined series, and they were used in about half of the pediatric series. Tonsillar resection was relatively common in the pediatric series, whereas shunting was common in the combined series.

Postoperative Outcomes: Syringomyelia

It is well established that CM-I is the leading cause of syringomyelia and that the creation (and worsening) of syringomyelia is one of the main causes of neurological symptoms and deficits in these patients. The improvement or resolution of syringomyelia is one of the main goals of the surgical treatment of patients with CM-I.[18,69,131] Improvement/resolution of syringomyelia was described in all of the reported series (pediatric, adult, combined, and total) for almost three-quarters of the patients (78%) (Fig. 8, Table 5), and there was no statistical difference in syrinx outcomes among any of the series subgroups.[1–9,11–19,21–63, 65–67,70,71,73–97,100,102–111,113–130,132–135,137–151,154–159] This outcome suggests that the use of operative decompression techniques achieved appropriate treatment goals in the majority of the patients. It should be noted, however, that about one-quarter of the patients (22%) still did not experience improvement. Therefore, additional means to further improve syrinx outcomes in this patient subgroup should be investigated.

Postoperative Outcomes: Neurological Status

Preoperative neurological deficits improved or resolved in 73% of the adult series, 84% of pediatric series, 72% of combined series, and 75% for all series (Table 6, Fig. 10). These satisfactory neurological results in approximately three-quarters of all patients correspond with the similarly good improvement/resolution of syrinx.[1–9,11–19,21–63,65–67,70,71, 73–97,100,102–111,113–130,132–135,137–151,154–159] Neurological improvement/resolution was better in the pediatric series, but the data did not reach statistical significance. However, the difference in the results of worsening neurological deficits in the follow-up period (7% vs 1%, adult vs pediatric, respec-

tively) did reach statistical significance. We may speculate that this difference is because younger patients frequently responded better to surgical decompression.

Postoperative Outcomes: Headaches

Suboccipital headaches aggravated by the Valsalva maneuver are one of the highlighted symptoms in all patients with CM-I.[1–9,11–17,19,21–63,65–67,70,71,73–97,100,102–111,113–130,132–135, 137–151,154–159] However, only 16% of these series reported postoperative headache outcomes. As the other aspects of postoperative results showed, the improvement or resolution of headaches was also good, occurring in 73% of adult patients, 88% of pediatric patients, and 81% in all patients (Fig. 11). Although good postoperative outcomes were shown in both adult and pediatric patients, the fact that the difference in improvement/resolution outcomes between pediatric and adult series did reach statistical significance further supports the hypothesis that pediatric patients have somewhat better clinical results after decompression. Nonetheless, these findings clearly and strongly support the indication for surgical decompression in patients of all ages who have incapacitating headaches as the sole symptom of CM-I.

Complications

Only 41% of the series reported complications,[1–9,11–19, 21–63,65–67,70,71,73–97,100,102–111,113–130,132–135,137–151,154–159] which is an interesting phenomenon. The median complication rate was 4.5%. It is unclear whether patients in the remaining 59% of the series did not have complications or simply did not report them. Of all series that reported complications, the most common were from the combined studies, followed by the pediatric and adult studies (Fig. 12).

Of all the pediatric series,[1–3,15,24,28,30–33,35,41,42,50,51,54,56,60,61, 65,77,80,81,86,89,92,93,97,104,108,109,116,133,134,143,145,150,154,155] 56% reported complications, with a median complication rate of 3.5%. The most common complications were pseudomeningocele, aseptic meningitis, CSF leak, meningitis, and neurological deficits (Fig. 13). The adult series that reported complications had a median complication rate of 3%, with CSF leak, aseptic meningitis, meningitis, and pseudomeningocele being the most frequent (Fig. 12). Clearly, CSF-related complications dominate in both pediatric and adult patient populations.

Postoperative mortality was rarely reported (only 11% of all series; 3% in pediatric series, 2% in adult series, and 3% in combined series),[1–9,11–19,21–63,65–67,70,71,73–97,100,102–111, 113–130,132–135,137–151,154–159] and there was no difference in the rates between adult and pediatric patients. The most common etiologies, in descending order of frequency, were pneumonia/respiratory failure, infection/sepsis, postoperative bleeding, and sleep apnea. The years of the series that reported complications were across the spectrum of the studied time interval. One may only speculate whether other series had no postoperative mortality whatsoever or if some series did not report it.

Conclusions

We have reviewed published operative CM-I series from the past 48 years and noted some novel, more precise information and provided propositions for future report-

ing of CM-I series. We have noted the increase in publications of surgical CM-I series in the past 21 years in all age groups. Most commonly, these series were published in the United States and Europe. The mean study duration was 10 years, and the mean follow-up time was 3.5 years. The median number of patients in the studies was 31.

Chiari I malformation was evenly distributed in adult and pediatric patients and was more common in female adults and children. In all the studies, the mean age was 27 years, and the median was 36 years. The peak ages of presentation were 8 years, followed by 9 years, in the pediatric series and 41 years, followed by 46 years, in the adult operative series. Two-thirds of all the patients in the operative CM-I series harbored syringomyelia, and its presence was more common in adults. In the adult and combined series, posterior fossa/foramen magnum decompression, dural opening/duraplasty, and arachnoid opening/dissection were used overwhelmingly. In pediatric patients, the frequency of arachnoid preservation versus arachnoid opening/dissection was evenly split.

The results of surgical treatment for patients with CM-I were excellent in all aspects: 78% improvement/resolution of syringomyelia, 75% neurological status improvement/resolution, and 81% suboccipital headache improvement/resolution. Postoperative headache outcome improvements were better in pediatric patients. A worsening of neurological status was more common in adult patients. Postoperative complications in pediatric patients were more commonly reported. Mortality is rare, most commonly resulting from pneumonia/respiratory failure, infection, sepsis, postoperative bleeding, and sleep apnea. For future publications on the subject of CM-I, we propose that authors provide details of all cases for the purpose of comparison, especially regarding the exact technique used.

Acknowledgments

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Author Contributions

Conception and design: KI Arnautovic, Splavski, Boop. Acquisition of data: all authors. Analysis and interpretation of data: all authors. Drafting the article: all authors. 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: KI Arnautovic. Statistical analysis: KI Arnautovic, Splavski, Boop. Administrative/technical/material support: all authors. Study supervision: KI Arnautovic, Splavski, Boop.

Correspondence

Kenan I. Arnautovic, 6325 Humphreys Blvd., Memphis, TN 38120. email: kenanarnaut@yahoo.com.