

## Chiari type-I malformation: Clinical features and surgical outcome assessment using Chicago Chiari Outcome Score (CCOS) among Sudanese patients

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### Abstract:

**Background:** Chiari malformation is one of the most controversial topics in neuro-surgery today. There is no of agreement as to what define these malformations, their symptoms and their natural history. Chicago Chiari Outcome Score (CCOS) is a scoring system designed to study the surgical outcome of Chiari type-1 patients.

**Purpose:** The purpose of the study aimed to assess patients with Chiari malformation type-1 in Sudan regarding their presenting symptoms, radiological findings, and surgical outcome for different surgical modalities and techniques.

**Material and Methods:** This study was a prospective observational study done at different neuro-surgery centers in Khartoum during the period from February 2018- August 2019. The study included all Sudanese patients who underwent decompression surgery for Chiari-I malformation with or without duraplasty during this period. Chicago Chiari Outcome Score (CCOS) was applied to evaluate post-surgical outcome.

**Results:** 20 patients were found to satisfy inclusive and exclusive criteria for Chiari-I malformation and were all operated (11 males and 9 females). The mean age was 33 years. The mean duration of symptoms was 19.8 months. Associated syringomyelia was detected in (18, 90%). All patients underwent posterior fossa decompression with or without removal of the posterior arch of C1, and with or without dura-plasty. Mean time for follow up was 4.45 months. Using the scoring system of CCOS, The outcome of the study group revealed in capacitated outcome (3, 15%), impaired outcome (6, 30%), functional outcome (9, 45%) and excellent outcome (2,10%). Patients without associated syringomyelia showed better outcome in comparison to patients with associated syringomyelia which was statistically significant (P = 0.008).

**Conclusion:** This study provides evidence that early surgical intervention for Chiari 1 malformation before the development of delayed sever symptoms of the disease was associated with better outcome especially in those without associated syringomyelia.

**Keywords:** Chiari malformation type-I, syringomyelia, post-operative outcome, Chicago Chiari Outcome Scale

### Introduction:

Chiari malformation is one of the most controversial topics in neuro-surgery today. Still there is no agreement as to define these malformations, their symptoms and their natural history. Therefore, a wide variety of techniques have been proposed for treatment of Chiari malformations.<sup>1</sup> Chiari malformation (CM) is a hindbrain disorder that is associated with de-

formity and elongation of the cerebellar tonsils. It is specifically characterized by the descent of the cerebellar tonsils >5 mm below the foramen magnum into the spinal canal.<sup>2</sup> CM type-I (CM-I) is the most common type. Although CM-I can occur in both pediatric and adult patients, the prevalence of CM-I in Sudan is not yet well defined. CM-I often becomes clinically apparent when the patient is aged 20–39 years. Thus,

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Table-1 : Chicago Chiari Outcome Scale<sup>10</sup>:

Pain	Non pain	Functionality	Complications	Total score
1. Pre-operative symptoms worse	1. Pre-operative symptoms worse	1. Unable to attend	1. Persistent complications poorly controlled	4. Incapacitated outcome
2. Un changed/refractory to meds/onset of poorly managed symptoms	2. Un changed/refractory to meds/onset of poorly managed symptoms	2. Moderate impairment (50%) attendance	2. Persistent complications well controlled	8. Impaired outcome
3. Improved/managed with meds/	3. Improved/managed with meds/	3. Mild impairment (>50%)	3. Transient complications	12. Functional outcome
4. Resolved/no onset of new symptoms	4. Resolved/no onset of new symptoms	4. Fully functional	4. Uncomplicated course	16. Excellent outcome

in the past, CM-I had been diagnosed primarily during adolescence or adulthood. However, with the advent of magnetic resonance imaging (MRI), the number of pediatric patients diagnosed with CM-I is increasing.<sup>3,4</sup> CM-I is usually associated with ventricular dilation, syringomyelia, cranio-cervical junction anomalies and scoliosis, but the relevant pathogenesis is not clearly understood. Moreover, the incidence of these conditions co-presenting with CM-I have varied considerably in the last recently published studies.<sup>2</sup> Neuro-surgical intervention plays an important role in the management of Chiari-I malformation in symptomatic patients, and the ultimate goal of surgery is to decompress the cranio-cervical junction to overcome the symptoms. There are several surgical options available for treating patients with Chiari-I malformation. This includes cranio-cervical decompression with or without duraplasty, 4<sup>th</sup> ventricular stenting, tonsillar reduction, and syringo-hydromyelia decompression.<sup>5-8</sup> Overall, symptoms related to Chiari-I malformation improve in 80–95% of cases after surgery. However, the complication rate reaches approximately 20% in adults and 37% in children, and the reported surgical mortality rate is around 1–11%.<sup>9</sup> A lot of researches and studies were done world wide regarding the pathophysiology of Chiari malformation type-1, the optimum surgical intervention and long term follow up after surgery. Some authors proposed cranio-cervical junction fixation. Chiari type-1 surgery is still debatable and challenging issue which needs more investigations and researches. One of the major limitations in Chiari researches was the lack of a standard outcome measure which could be utilized as the basis in a variety of researches for the purpose of com-

paring different management and surgical techniques besides comparing the results of the different studies against each other.

Chicago Chiari Outcome Score (CCOS) is a scoring system designed to study the outcome of Chiari patients. Applying CCOS scoring to a larger patient population undergoing variety of operative CM-1 techniques should allow for better-informed decisions regarding patient selection and treatment options for patients with CM-1.<sup>10</sup> The CCOS is composed of 4 main 3 categories: Pain, Non-Pain symptoms, Functionality, and complications. A patient is given a score of 1-4 in each category, for a total score of 4-16. A final score of 4 means the person is incapacitated; a score of 8 means they have an impaired outcome; a score of 12 equates to a functional outcome; and a score of 16 is of course an excellent outcome (table-1).<sup>10</sup> In Sudan, Chiari malformation type-1 is one of the frequently seen neuro-surgical diseases, but unfortunately there is no documented data available or a computerized patient registry system. Therefore, the aim of this study is to establish a future database for Chiari malformation type-1 regarding the presenting symptoms, radiological findings and surgical outcome for different surgical modalities and techniques. Also this study is aiming at reviewing the surgical outcome of Chiari malformation type-1 using Chicago Chiari Outcome Score for patients after surgery, and to correlate the different types of surgical intervention done with the outcome besides describing and justifying post-operative complications for each surgical technique.

Table-2 Presenting symptoms among study group:

Symptoms	Number of Patients	Percent%
Valsalva headache	18	90
Blurring of vision	9	45
Vertigo	5	25
Difficult swallowing	6	30
Poor sleep	13	65
Fatigue –day time sleep	17	85
Balance disturbance	10	50
Speech disturbance	6	30
Neck pain	18	90
Back pain	16	80
Arm pain	17	85
Limbs numbness	19	95
Limbs weakness	14	70

Table-3 Surgical categories performed among study group:

Surgery type	Frequency	Percent
Sub-occipital craniectomy	4	20.0
Sub-occipital craniectomy +c1 posterior arch removal	4	20.0
Sub-occipital craniectomy +c1 posterior arch removal +duraplasty	10	50.0
Sub-occipital craniectomy +c1 posterior arch removal +c2 posterior arch removal +duraplasty+ syrinx opening through myelotomy	1	5.0
Suboccipital craniectomy +c1 posterior arch removal +c2 posterior arch removal +duraplasty + syringe-subarachnoid shunt	1	5.0
Total	20	100.0

Table-4 Pre-and post-operative comparison of symptoms and signs among the study group:

Symptoms	Pre-operative s	Post – operative symptoms		
	Number of Patients + Percent%	Resolved	Improved	Remained the same or got worse
Valsalva headache	18/20=90%	18/18 =100%	-	-
Blurring of vision	9/20=45%	7/9=7.8%	2/9=22.2%	-
Vertigo	5/20=25%	5/5=100%	-	-
Difficult swallowing	6/20=30%	5/6=83.3%	1/6=16.7%	-
Poor sleep	13/20=65%	7/13=53.8%	5/13=38.5%	1/13=7.7%
Fatigue –day time sleep	17/20=85%	11/17=64.7%	4/17=23.5%	2/17=11.8%
Balance disturbance	10/20=50%	5/10=50%	2/10=20%	3/10=30%
Speech disturbance	6/20=30%	3/6=50%	3/6=50%	-
Neck pain	18/20=90%	15/18=83.3%	2/18=11.1%	1/18=5.6%
Back pain	16/20=80%	12/16=75%	2/16=12.5%	2/16=12.5%
Arm pain	17/20=85%	12/17=70.6%	3/17=17.6%	2/17=11.8%
Limbs numbness	19/20=95%	10/19=52.6%	4/19=21.1%	5/19=26.3%
Limbs weakness	14/20=70%	2/14=14.3%	5/14=35.7%	7/14=50%

## Material and Methods:

This study was a prospective observational study done at different neuro-surgery centers in Khartoum during the period from February 2018-August 2019. The study included all Sudanese patients who satisfy clinical and radiological features of Chiari-1 malformation and underwent surgical decompression with or without dura-plasty. Inclusive criteria include all patients presented with the clinical and radiological features of the disease and who were operated in one of these centers (NCNS, Neuro-spine Center-Ribat Teaching Hospital, Military Hospital, and Omdurman Teaching Hospital). All patients who did not come for regular follow up post-surgery or operated in centers other than the aforementioned centers were excluded from the study. Data was collected using a questionnaire investigating the pre and post operative symptoms, signs, radiological findings. Chicago Chiari Outcome Score (CCOS) was applied to all cases in the study to evaluate post-operative outcome. All patients included in this study had been consented to share in this study by a written informed consent. Data confidentiality maintained using the indexing methods to insure that. Data was then collected in a designed SPSS sheet and analyzed using SPSS version 20.

## Results:

20 patients were included in this study (11 males and 9 females) (figure-1). The mean age at presentation was 33.45 years. The presenting symptoms are summarized in (table-2). The symptoms durations are shown in (figure-2) with mean duration of 19.8 months. Syringomyelia was seen in 18 patients (18/20, 90%). The syringomyelia seen in cervical spine in 9 patients (9/18, 50%), and in cervico-dorsal spine in 9 patients (9/18, 50%). All patients underwent cranio-cervical decompression surgery with or without removal of the posterior arch of C1, C2, and with or without dura-plasty (table 3). CSF leak was the most common post-operative complication and was seen in 9 patients (9/20, 45%). Hydrocephalus was seen in 2 patients as one of the complications.

Post-operative outcome: The headache was

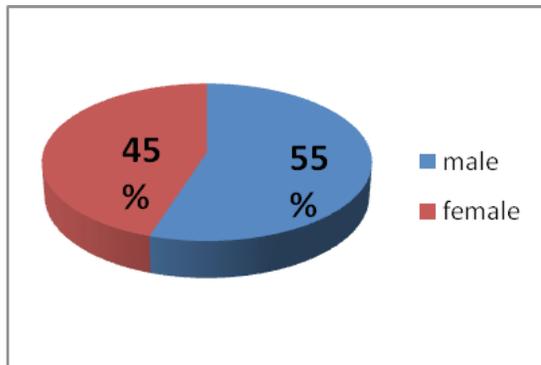


Figure 1: Distribution of sex among study group

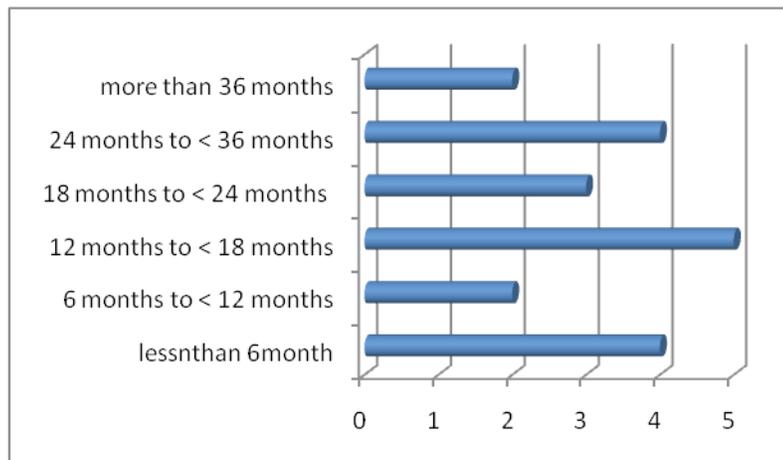


Figure 2: Symptom duration among study group

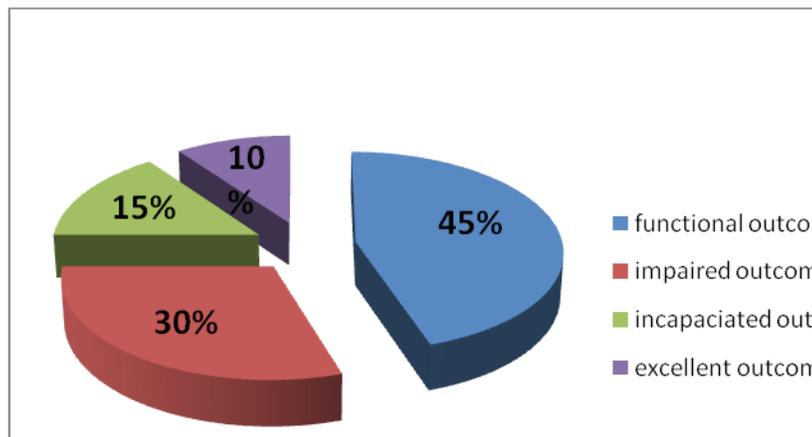


Figure 3: CCOS among study group

subsidised in all patients with pre-operative headache. The weakness resolved in 2-patients (2/14,14.3%) and improved in 5-patients (5/14, 35.7%) while in 7-patients (7/14, 50%) the weakness remained the same or got worse. table-4 demonstrates the comparison between pre-operative symptoms of patients in this study and the outcome post-operatively. Mean time

for follow up was 4.45 months (the range from 2 months to one year). Using the scoring system of CCOS: as shown in figure (3); the outcome of the study group revealed incapacitated outcome in 3 patients (3/20, 15%), impaired outcome in 6 patients (6/20, 30%), functional outcome in 9 patients (9/20, 45%), excellent outcome in 2 patients (2/20, 10%). No significant differences in outcome between males and females (p value = 0.8). Patients without syringomyelia showed better outcome than patients with syringomyelia which was statistically significant (P = 0.008). Patients underwent early surgery showed better outcome. Also patients showed better outcome with sub-occipital craniectomy + C1+ duraplasty than other types of surgery. Syringomyelia extension in the cord also plays a role in the outcome (i.e. outcome in patients with cervical syringomyelia was found better than outcome in patients with cervico-dorsal syringomyelia. Examples of pre-operative, intra-operative, and post-operative MRI images and photo are shown in the illustrations (1) & (2).

**Discussion:**

Chiari malformations have been considered as rare conditions, but with the introduction of MRI, Chiari malformation type-I is more commonly diagnosed with a prevalence of 0.5%-3.5% as reported by the recent studies.<sup>1</sup> Patients included in this study present with wide range of symptom varieties ranged from headache to myelopathy symptoms. All of the symptoms associated with CM-1 in this study are also demonstrated by previous studies done worldwide with close percentages.<sup>11-14</sup> The duration of symptoms before surgery has been found to affect the outcome. This may be one of the factors that explain the poor outcome in this study, because patients with longer duration of pre-operative symptoms showed less improvement when compared to patients with short pre-operative duration period. This fact was also reported before by many authors in the literature.<sup>1,13-15</sup> Syringomyelia was seen in most of the patients in this study group. This was higher than any previously reported study and this may be attributed to the late presentation and diagnosis in our



Illustration 1: of one of the patients in the study group. left side pre-operative MRI , note the tonsillar herniation and syringomyelia. Right side post-operative MRI of the same patient showed hydrocephalus as a complication, note the decompression site



Illustration 2: of one of the patients in the study group. left side pre-operative MRI , note the tonsillar herniation and syringomyelia. Right side intra-operative picture of the same patient showed herniated tonsils after bony decompression and dural opening.

study group. Platybasia and scoliosis was seen as association with Chiari MF-1 in some patients in our study group but with less frequency than in what is reported before in the literature. All patients underwent decompression surgery with or without dura-plasty. CSF leak was the most common post-operative complication encountered. Also meningitis and hydrocephalus were seen in few cases of our study group. These findings correspond with the previous studies.<sup>1,16-18</sup> The patients were evaluated for their symptoms, daily activities and return to their initial jobs and functions using the scoring system of CCOS. Almost half of the patients in the study showed functional outcome, while only few of them showed excellent outcome. This may be partly explained by the delayed presentation and the presence of associated syringomyelia in most of the cases. The outcome of this study was close to

the surgical outcome reported by Nkusi et al in Rwanda.<sup>11</sup> In this study, measurements for syringomyelia was not done neither before nor after surgery, but syringomyelia opening was done in 2-patients one through Myelotomy and one through syringo-sub-arachnoid shunt and both did not improve post-operatively. Syringomyelia extension affects outcome. Outcome in patients with cervical syringomyelia was better than outcome in patients with cervical and dorsal syringomyelia. The outcome of Chiari surgery with many surgical procedures is good in the first few months which then disappear over time. Whether this is due to a placebo effect or something else, this may reflect the fact that after a period of time, most Chiari patients have learned to manage their residual symptoms through lifestyle modifications and have mentally accepted that they will likely never feel they will be 100% fine. It is this adaptation and accommodation which is often not adequately captured in the current outcome researchs.<sup>19</sup> Increasing the awareness of patients and doctors in the different disciplines about Chiari Malformation symptoms, signs and diagnosis may help in the early presentation, diagnosis and consequently, early surgical treatment and better outcome.

Lack of follow up post-surgery is one of the major limitation in this study because most of the patients reside far away from the neuro-surgical centers and need to travel several miles before reaching these centers. The other limitations include lack of neuro-surgical and diagnostic facilities in the remote states of Sudan and lack of good documentation of the patient's clinical and operative data. This study represents one of the few studies about the quantitative post-operative outcome of Chiari type-1 in the region and the first one in Sudan. However, further studies with a larger number of patients and a longer duration are needed to better assess the outcome and the future trends and guidelines for treating patients with Chiari-I malformations.

#### Conclusion:

The delay in diagnosing and managing CM-I patients can affect the surgical decision and outcome. Increasing the awareness of doctors and

health care providers about Chiari symptoms, signs and diagnosis may lead to early presentation and diagnosis then early surgical treatment to achieve a better outcome. Adopting a quantitative measure to assess the surgical techniques and post-operative outcome is the future for Chiari Type-I researches. This will allow not only in comparing what is the most appropriate technique and what is best for the patient, but will also allow for comparing the studies against each other. This can be achieved through using the existing scales like Chicago Chiari Outcome Scale or through creating new scales based on the natural history of the disease in the region.

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**Role and contribution of authors:**

Dr Yasser Seddeg Abdulghani, collected the references, data and did the initial writeup.

Dr Mohammed Awad Elzain, collected the references and helped in collecting the data and also improved the initial writeup.

Dr Ali Awad Ali, helped in collecting the data and also helped in introduction writing.

Dr Haytham Hussien Mohamed, helped in collecting the references and also helped in discussion writing.

Dr Basheer Mohammed Basheer, critically review the article and made the final changes.

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