

Surgical Outcome of Chiari
Malformations Type 1 in RwandaNkusi AE^{1,2}, Muneza S^{1,2}, Hakizimana D^{1*}, Nshuti S² and Munyemana P²¹Department of Surgery, University of Rwanda, Rwanda²Department of Surgery, University Teaching Hospital of Kigali, Rwanda

Article Information

Received date: Oct 29, 2017

Accepted date: Jan 05, 2018

Published date: Jan 09, 2018

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Keywords Chiari; Suboccipital; Craniectomy; Duroplasty; Pericranial Flap; Outcome

Abbreviations MRI: Magnetic Resonance Imaging; CSF: Cerebrospinal Fluid; CM-I: Chiari Malformation type I; PICA: Posterior Inferior Cerebellar Artery; BAE: Brainstem Auditory Evoked potential; MEPS: Motor-Evoked Potentials; SEPs: Somatosensory-Evoked Potentials

Abstract

Background: Chiari Malformations are a collection of hindbrain abnormalities ranging from simple herniation of the cerebellar tonsils through the foramen magnum to complete agenesis of the cerebellum. Symptoms of Chiari I develop as a result of pathophysiological consequences of the disordered anatomy. Early surgery is recommended for symptomatic patients but the most effective surgical technique is a matter of controversy.

Methods: This study is an observational retrospective study on surgical outcome of all patients operated on for Chiari malformation type 1 from January 2010 to May 2015 in the two tertiary referral hospitals of Rwanda.

Results: Twenty one patients were enrolled in the study, 11 males and 10 females with a mean age of 37.3 years. The most commonly performed procedure in our study for Chiari malformation type I involved suboccipital craniectomy with resection of the posterior arch of atlas, coagulation of herniated cerebellar tonsils and duroplasty. At 6 months follow up 90% of our patients had moderate to very much improvement, 10% had slight improvement and no one reported worsening or surgery related complications.

Discussion and conclusion: This surgical series of Chiari malformation type I patients comes as one of the rare series of its kind from African continent and it is illustrating the effectiveness and safety of well selected posterior fossa decompression with autologous pericranial graft duroplasty in this era of controversy regarding whether or not duroplasty should be added to posterior fossa decompression in the treatment of Chiari malformation type I.

Background

Chiari malformations are a collection of hindbrain abnormalities ranging from simple herniation of the cerebellar tonsils through the foramen magnum to complete agenesis of the cerebellum. They are referred to as Chiari malformation after Dr. Hans Chiari who in 1891 used autopsy specimens to describe four types of congenital anomalies of the hindbrain [1].

The Chiari Malformation type I (CM-I) is the most common and the least severe amongst other types, often diagnosed in adulthood. It consists of caudal displacement of the cerebellar tonsils into the upper cervical spinal canal. The most common associated radiological findings are cervical syringomyelia and hydrocephalus in some occasions [2].

The pathogenesis of Chiari malformations has been passionately debated since the initial descriptions by Hans Chiari, observations that remain as valid today as they were more than a century ago. Different hypotheses have been proposed and mostly based upon mechanical events, e.g: crowding theory, pulsation theory, traction theory, hydrodynamic oligo-cerebrospinal fluid theory. None satisfactorily addresses all features of the Chiari malformations [3].

A molecular genetics hypothesis was proposed as well to try to explain not only the shallow posterior fossa but also intrinsic focal intradural pathologies, and myelodysplasia including meningomyelocele and segmental hydromyelia. The molecular theory explains that the Chiari malformations are primary defects in the genetic programming of hindbrain segmentation and of growth of associated structures of the chondrocranium [4].

Symptoms of Chiari I develop as a result of 3 pathophysiological consequences of the disordered anatomy: compression of medulla and upper spinal cord, cerebellum infarct from compression of the Posterior Inferior Cerebellar Artery (PICA), and disruption of CSF flow through foramen magnum. Compression of cord and medulla may result in myelopathy, lower cranial nerves dysfunction. PICA compression may result in vertigo, nausea and truncal ataxia. Disruption of CSF flow through foramen magnum probably accounts for the most common symptom; pain.

The MRI is the diagnostic imaging modality of choice in symptomatic patients. But the criteria for the descent of cerebellar tonsils tip below the foramen magnum to diagnose the Chiari I malformation is controversial and requires clinical correlation.

Early surgery is recommended for symptomatic patients and surgical options include: suboccipital craniectomy with C1 laminectomy and band resection with or without cerebellar tonsils coagulation and duroplasty.

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ISSN: 2573-6728

Table 1: Clinical presentation.

Symptoms/signs	Frequency	Percentage
Headache	14	66.70%
Neck pain	13	61.90%
Myelopathy	18	85.70%
Upper limb numbness	8	40%
Upper limb weakness	15	65%
Spastic gait	7	35%
loss of balance	1	5%
Urinary urgency	1	5%
Urinary incontinence	0	0%
Urinary retention	1	5%
Cerebellar symptoms	4	19.05%
Limbs ataxia	2	11.10%
Dysarthria	1	5%
Dysphagia	1	5%
Down beat nystagmus	2	11.10%
Central cord syndrome	2	9.52%

This article is focusing on clinical and radiological presentations, surgical treatment and its outcome; of Chiari malformation type 1.

Patients and Methods

This study is an observational retrospective study of all patients operated on for Chiari malformation type 1 from January 2010 to May 2015 in the two tertiary referral hospitals of Rwanda which neurosurgical services. Eligible patients were identified from the patients’ registries of the neurosurgery unit, operating room, outpatients department and radiology department of University Teaching Hospital of Kigali and King Faisal Hospital, Rwanda. The patients’ files were consulted to verify the date of diagnosis and to make sure that the identified patients fit the inclusion and exclusion criteria. Patients were called and seen in outpatients department and

fill the questionnaire that was used to collect the data on the post-operative outcome. A questionnaire was used to record the following data: patient’s age, gender, district of residence, presenting symptoms and signs, radiological findings, the used surgical technique and patient reported outcome 6 months after surgery.

Results

Twenty eight patients were operated for Chiari malformation type I during the study period. Seven of them were excluded from analysis because of incomplete data. Twenty one patients were included in the analysis. Male were 11 (52.38%) and females were 10 (47.62%). The mean age of our study population was 37.3 years, the youngest patient was 19 years old, and the oldest one was 72 years old.

Headache was the most common presenting symptoms (66.7%), neck pain was the second frequently reported symptom (61.9%). Sixty percent of our patients had upper limb weakness, 40% had upper limb numbness and cerebellar dysfunction was present in 19% of cases (Table 1).

Brain and /or cervical spine MRI was used for diagnosis in all cases. The mean length of cerebellar tonsils descent below the level of foramen magnum on MRI sagittal scan was 9.3 mm (SD 2.6). Twelve patients (57%) had syrinx.

All 21 patients underwent posterior fossa decompression that involved a standard suboccipital craniectomy with or without additional surgical procedure steps (Figure 1). The most commonly performed procedure in our study for Chiari malformation type I involved suboccipital craniectomy with resection of the posterior arch of atlas, coagulation of herniated cerebellar tonsils and duroplasty. This was done for 14 patients (66.7%). Four patients (19%) underwent suboccipital craniectomy combined with resection of the posterior arch of atlas and autologous graft duraplasty without reducing the size of herniated tonsils. In total, dura opening and augmentation with an autologous pericranial graft was performed in 85.7 % of cases (n=18). Three patients underwent extradural posterior fossa decompression alone.

In this series we did not record any case of death. None of our patients had CSF leak, pseudomeningocele or neurological deterioration after surgery.

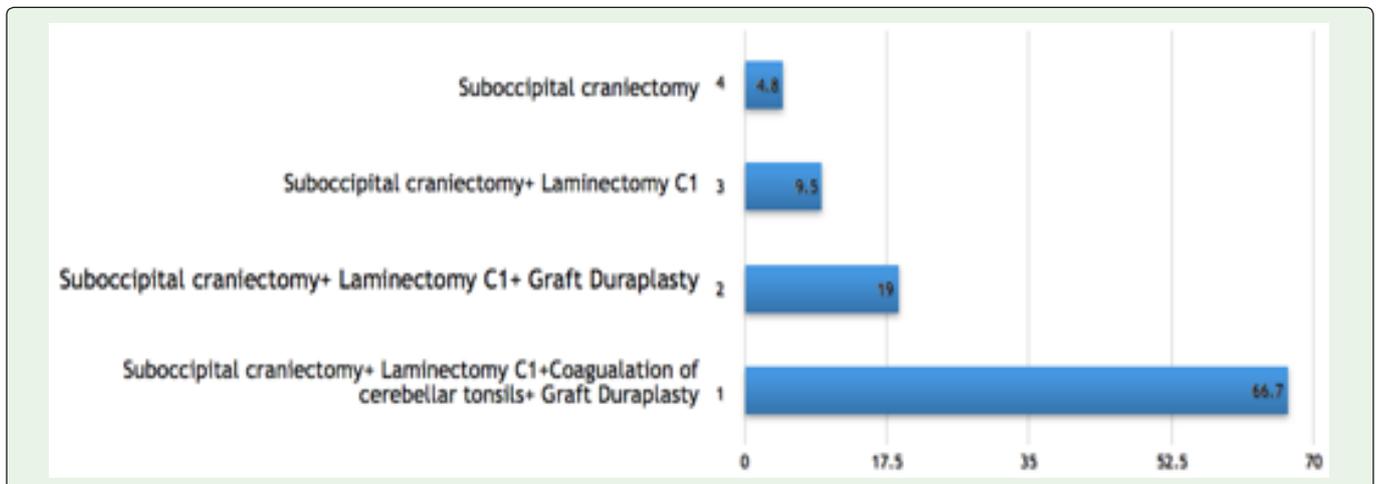
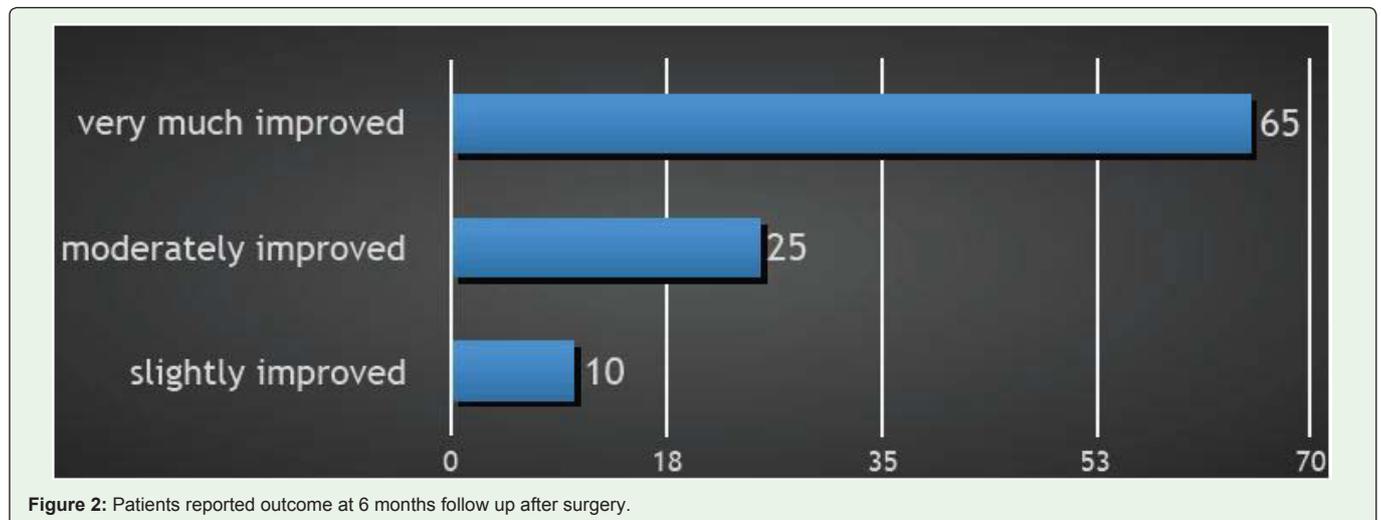


Figure 1: Performed surgical procedures.



Euro-Qol-5D [EQ-5D] was used as a tool to qualitatively measure the outcome after surgery. It is a 5 item questionnaire where the patient was asked to mark an item that was best reflecting her or his quality of life at 6 months after surgery compared to the her or his condition before surgery.

Sixty five percent of our patients responded to have improved very much, 25% had a moderate improvement and 10% responded that they had experienced a slight improvement (Figure 2).

Patients who presented with headache tend to improve better than those without headache, 78.5% vs. 28.5% ($p = 0.02$).

Discussion

Chiari malformations have been considered as rare conditions, but with the arrival of MRI, Chiari malformation type I is more commonly diagnosed with a prevalence of 0.5%-3.5 % reported by recent studies [5,6]. This surgical series of Chiari malformation type I patients comes as one of the rare series of its kind from African continent as it was illustrated by one review of surgical series of Chiari malformation type 1 that found only 1 publication from Africa among 145 operative series of Chiari malformation type 1 reported in English literature until 2013, the majority of them came from North America and Europe. The mean size of these series was 31 patients over a mean period of 10 years. Our study included a similar number of patients ($n = 28$) over less than a half of the above mentioned period of time (4.5 years). This may be explained by systematic use of MRI and a high suspicion index of Chiari malformation in patients with cervicogenic headaches. The mean age of our study population was 37.3 years, which is similar to the one reported by Aska Alamovic et al (mean age was 33) in their literature review of surgical series published between 1965–2013 on pediatric and adult Chiari malformation Type I. The association of syringomyelia with CM-I has been reported to range from 37% to 75% [7-9]. The prevalence of syrinx in our study population (57%) fell into that range as well. The outcome of the surgical treatment is excellent in most of the cases (78%) if surgery is performed within 2 years of symptoms onset [10].

As patients with Chiari malformation type I may present with a variety of symptoms and signs ranging from headache to severe myelopathy and brainstem dysfunction, this condition may mimic a

wide range of neurological conditions. The diagnosis requires high level of awareness of its presentation, availability and a systematic and clinically oriented analysis of the MRI of the craniocervical junction. Once the diagnosis is made the patient and patient's family expect to get from the surgeon the information about available surgical options, their associated complications and their outcome in terms of improvement and prevention of worsening of patient's symptoms. In addition, the surgeon faces a challenge of selecting the best surgical procedure for an individual patient, both pre and intra-operatively. In order to overcome to these challenges the surgeon has to be armoured with the data from his/her surgical series in comparison with those from other centres' series reported in the literature.

Even if it is currently well established that surgery is the best treatment for Chiari malformation type 1, it is still a subject of controversy what is better between posterior fossa decompression with duroplasty and posterior fossa decompression without duroplasty. Posterior fossa decompression with duroplasty has been demonstrated to yield to better clinical improvement and lower rate of re-operation [11,12]. The drawbacks of posterior fossa decompression with duroplasty are higher overall complications rate and CSF related complications rate [13]. In our study, the extent of posterior fossa decompression was based on intra-operative assessment of adequate CSF flow through the foramen magnum and obex that was confirmed by CSF pulsation. By doing so, posterior fossa decompression with autologous pericranial duroplasty was performed in 85.7 % and coagulation/resection of herniated cerebellar tonsils was done in 66.7% of cases. There was no death, no surgery related neurological injury and no CSF related complication in our patients. 90% of patients reported moderate to very much improvement, 10% had slight improvement and no one reported worsening. Similar results were reported by L. Zhang et al in their study that compared autologous and synthetic graft duroplasty among 27 patients with symptomatic Chiari malformation type I [14]. In that study all 27 patients demonstrated improvement of preoperative symptoms and good wound healing. As comparison, among 4206 patients included in surgical series reviewed by Aska Alamovic et al, where arachnoid opening and dissection was performed in 70% of cases and tonsils size reduction done in 23 % of cases, 75% of patients reported to be improved, 16.6% had no change in neurological status, and 8.5%

experienced worsening. This difference in outcome depending on how much the posterior fossa volume was increased surgically was demonstrated by other studies [15]. Another point of controversy entails what should be the criteria to be based on to judge if addition of duroplasty to extradural posterior fossa decompression is required or not. Some authors considered the presence of syringomyelia and previous posterior fossa decompression surgery as indications of duroplasty and others have advocated the use intraoperative BAEs conduction changes, MEPs and SEEPS amplitudes changes as a guide to predict which patients will benefit from suboccipital craniectomy with or without duroplasty [13,16,17]. But till now there are no high quality data to support any of these methods [18].

Conclusion

This operative case series demonstrates that Chiari malformation type I exist in Africa like in any other place in the world. With wider availability of MRI and neurosurgeons, more Chiari malformations type I are going to be recognised and effectively treated. Good results are almost uniformly obtained with adequate posterior fossa decompression which restores normal CSF flow. However, the indication of intradural or extradural decompression for a particular patient is left to surgeon's discretion and remains a subject of debate. Randomised clinical trials are warranted to provide strong evidence regarding indications of posterior fossa decompression with or without duroplasty.

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