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The Seow Operative Score (SOS) as a decision-making adjunct for paediatric Chiari I malformation: a preliminary study

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Abstract

Purpose The natural history of Chiari I malformation (C1M) in the paediatric population is poorly understood. There are conflicting reports with regards to surgical indications, operative techniques and peri-operative prognostic indicators. In this paper, we report our institutional experience in the management of paediatric C1M.

Methods The workflow process which includes preferred imaging modalities, surgical techniques and indications for intervention is discussed. In particular, we describe one of our current projects—an in-house designed Seow Operative Score (SOS) as a feasibility scoring system for neurosurgical intervention in our local cohort of paediatric C1M patients.

Results In our series, we have 2 groups: 10 non-operated patients versus 19 operated patients. In the non-operated group, the majority of patients had a SOS of 0 to 1. One patient had a score of 2.5 and was kept under close surveillance. Follow-up imaging demonstrated resolution of the cerebellar herniation and intraspinal syrinx. In the operated group, 17 patients had a SOS of 3 or more. Two patients had a SOS of 2. For these 2, 1 developed progressive symptoms, and the other had an extensive cervicothoracic syrinx. Decision was made for surgery after a period of surveillance.

Conclusions In this paper, we report our institutional experience in managing paediatric C1M and, at the same time, highlight salient points of our practices. Meanwhile, we advocate collective global efforts and in-depth research for better disease understanding of this challenging condition.

Keywords Chiari I malformation · Suboccipital decompression

Introduction

Chiari I malformation type I (C1M) is a heterogenous group of hindbrain disorders characterised by elongation of the cerebellar tonsils, which descend below the foramen magnum into the spinal canal [1]. Radiologically, it is defined as cerebellar tonsillar descent of 5 mm or more beyond the foramen magnum [1]. Although it was first described in the nineteenth century [2], the natural history of C1M, especially in the paediatric population, remains poorly understood [3, 4]. Existing literature has conflicting reports with regards to results of posterior fossa decompression variations, patient outcomes and peri-operative prognostic indicators [4, 5]. Atypical presentations of C1M may also cause diagnosis to be delayed [6–11]. Furthermore, certain conditions have been observed to be particularly associated with C1M in the paediatric cohort—these include growth hormone deficiency, scoliosis, congenital syndromes and central nervous system (CNS) infection [12–15].

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Paediatric Chiari I malformation in KK Women's and Children's Hospital: an overview

Our Neurosurgical Service is no stranger to the challenges faced in the management of children who present with this perplexing condition. In this paper, we report our institutional experience in the management of paediatric C1M. The majority of our cases are inter-departmental referrals whereby the diagnosis of C1M is based on there is cerebellar ectopia

visualised on magnetic resonance imaging (MRI). For ease of auditing patient care and outcomes, our unit has a customised workflow the moment they are referred to us.

As standard practice, MRI studies of the brain and whole spine are performed to assess for hydrocephalus, spinal syrinx and other related intracranial pathology. In recent years, we included the use of cine phase-contrast MRI to examine CSF flow characteristics caused by crowding at the foramen magnum. However, some patients forego this investigation due to cost issues and/or they are wearing orthodontic braces which cause too much artefacts in the MRI sequence. Following that, we assess the bony stability and integrity of the cranio-cervical junction with a flexion/extension X-ray of the cervical spine. This is based on previous claims that the sequalae of C1M are possible secondary effects of primary, longstanding atlantoaxial instability [16]. At times, a computed tomographic (CT) cervical spine scan may be done for selected patients if aberrant anatomy is strongly suspected (Fig. 1). The patient is then assessed with a thorough history and clinical examination, in conjunction with all his/her radiological results. Overall, we believe that the approach to paediatric C1M should be holistic, and it relies on three inter-dependent factors: radiology, patient symptoms and contributing medical problems.

The Seow Operative Score (SOS): a decision-making adjunct for surgery

Decision to intervene is often contentious for C1M patients. Although indications for surgery are known to vary among neurosurgeons, the general agreement is to offer treatment patients with a syrinx, in the face of progressive neurological symptoms [4]. Nonetheless, other complaints experienced by C1M patients tend to be more subjective and less well-articulated by the paediatric population. Under such circumstances, symptoms such as headaches, dizziness, visual disturbances and so forth may too be only manifested by crying and irritability.

In order to address these issues, we designed an in-house scoring system (SOS) to assess the feasibility of neurosurgical intervention in patients with a diagnosis of C1M. Using a modified propensity score modelling approach, multiple variables based on radiological, clinical symptoms and underlying medical conditions were curated from our patient cohort. These findings were corroborated with associated factors reported in paediatric C1M from the literature. Logistic regression was applied to internally validate our results. The SOS scores were divided into three categories according to a point system: '*Probable*' (3 points or more), '*Possible*' (2 to 2.5 points) and '*Unlikely*' (less than 2 points). Cases which fall under the '*Possible*' category are kept on close surveillance. More importantly, the disease expectations of such patients are discussed at length with their caregivers before and during this period of monitoring (Table 1).

At this point, we emphasise that this SOS scoring system is an ongoing work in progress. Currently, it serves only as an ancillary guide and it is no means absolute. Final decision-making for surgery remains dependent on patient factors and a consensus after discussion of each case at departmental level.

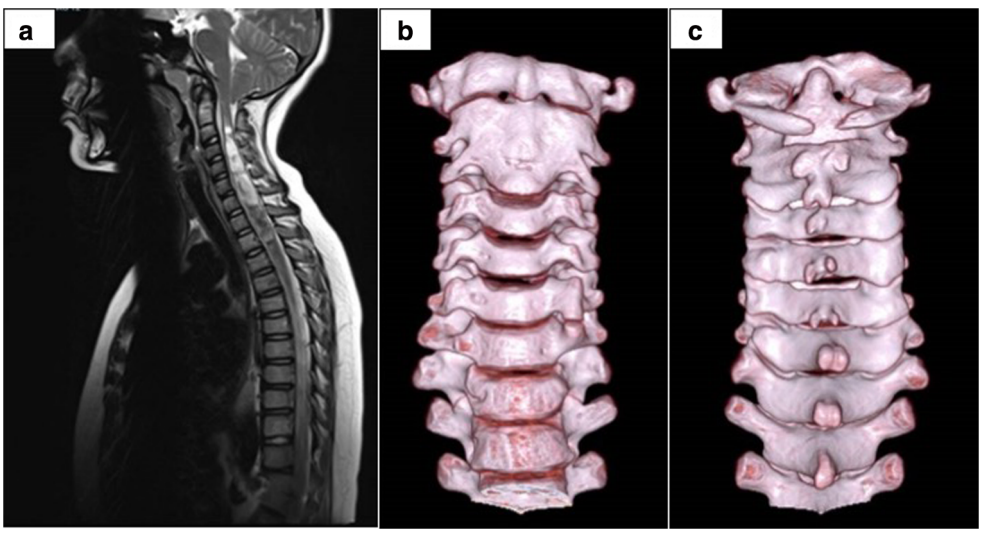

Fig. 1 **a** T2-weighted sagittal MRI image of an 11-year-old male with a background of Goldenhar syndrome. There are low-lying cerebellar tonsils and a cervico-thoracic syrinx. Decision was made for bony imaging of his cervical spine to aid surgical planning. **b**, **c** The three-dimensional reconstruction of his cervical spine in anterior and posterior views, respectively. There is non-fusion of the posterior arch of C1, and congenital fusion of C2-C3 vertebrae as well as the spinous processes, with rudimentary hypoplastic C2-C3 intervertebral disc. Incidentally, the odontoid peg appears slightly hypoplastic

Suboccipital decompression and duraplasty—our rationale

All the neurosurgeons in our unit use a similar surgical technique for C1M—suboccipital decompression, partial removal of the posterior arch of C1 and duraplasty. The main objectives for this approach are firstly the creation of corporal space to ease crowding at the foramen magnum and the restoration of adequate CSF flow in the region. Essentially, we believe that there are two perpetuating components in symptomatic C1M. These include *structural* (that is, bony and tissue elements that physically contribute to the crowding in the foramen magnum) and *dynamic* (here, the impedance of temporal CSF flow in the cranio-cervical junction) factors.

Additional nuances of our approach include adhesiolysis on thickened arachnoid with sharp dissection using micro-instruments. Also, the inferior edges of the cerebellar tonsils are gently cauterised in pulses to shrink them. Excision of the tonsils is usually avoided to decrease the risk of bleeding into the surgical cavity, which may then lead to new adhesions. Similarly, constant effort is made intra-operatively to wash out all unwanted surgical by-products from the operating site. Duraplasty is routinely done with a generous bovine graft in a watertight fashion. We are of the opinion that a graft has several advantages. First and foremost, it protects the underlying neural parenchyma from chemical contamination. Next, it creates a capacious sac to optimise the bony decompression. Last, but not least, it allows more intradural space for CSF circulation; hence, reducing possibility of post-operative adhesions.

Table 1 The Seow Operative Score (SOS) is calculated via a point system utilizing variables based on radiological, clinical symptoms, and underlying medical conditions. Cumulative scores were divided into three categories according to a point system: '*Probable*' (3 points or more), '*Possible*' (2 to 2.5 points) and '*Unlikely*' (less than 2 points)

Radiology	
Syrinx	1
HCP	1
Cine CSF flow disturbance	0.5
Associated conditions	
Progressive scoliosis	1
Syndromic/ Craniosynostosis	1
Previous CNS Infection	1
GH deficiency	1
Symptoms	
Sleep apnoea	1
Cranial nerve palsy(s)	1
Long tract signs	1
Tussive headache	0.5
TOTAL	**10**

Preliminary results and focused discussion

In our series, we have 2 groups: non-operated (10 patients) versus operated (19 patients) for children diagnosed with C1M. All these patients were managed by our unit from 01 January 1997 to 28 February 2019. Patients with basilar invagination, atlantoaxial dislocation and space-occupying lesion in the cranial fossa were excluded. For the cohort that did not undergo surgery, there were five males and five females (Table 2). The mean age of C1M diagnosis was 8 years old (youngest 1 year old, oldest 13 years old), and the average level of cerebellar tonsillar descent was 9.1 mm. Two patients in this group had syrinxes (patients 1n and 2n); one of them demonstrated spontaneous regression of her syrinx on an interval MRI. Interestingly, the same MRI also showed the resolution of her initial cerebellar tonsil herniation (patient 1n; Fig. 2). At the time of writing this manuscript, the remaining patient (patient 2n) was still asymptomatic and awaiting her follow-up MRI.

In the operated cohort, there were 7 males and 13 females (Table 3). The mean age of diagnosis was 9.85 years (youngest 1 year old, oldest 18 years old) and the average level of cerebellar tonsillar descent was 10.25 mm. The median length of stay after surgery was 5.95 days (shortest 3 days, longest 14 days). Scoliosis was evident in 13 patients, and 6 of them presented with progressive scoliosis. Also, three patients had a background of growth hormone (GH) deficiency and five patients were syndromic. In the latter subgroup, a patient (patient 4o) with Beare-Stevenson cutis gyrate syndrome had to undergo two interval surgeries: at 1 year and at 5 years old. Of note, one of the patients (patient 10o) with GH deficiency was a triplet child, and his sibling had craniosynostosis. Magnetic resonance imaging was performed as part of the workup for GH deficiency. This demonstrated low-lying cerebellar tonsils and an extensive cervico-thoracic syrinx. As he was asymptomatic, his SOS was 2. Decision was made for surgery after discussion with his parents. Following that, another patient (patient 17o) with a SOS of 2 was kept on regular follow-up. However, during this period of observation, she developed complaints reminiscent of sleep apnoea. After surgery, her symptoms improved significantly. As for our complications, one patient developed a pseudomeningocoele (patient 14o) which required surgical repair, whilst another had superficial wound dehiscence (patient 19o) that was managed conservatively. Both have remained well. No complications of iatrogenic CNS infection, neurological worsening, or death were encountered. All patients undergo a follow-up MRI

Table 2 Table showing list of C1M patients who were in the non-operated (hence, labelled as 'n') group. Under the heading 'Symptoms', 'Incidental' implies that the radiological C1M findings were incidental. These patients did not complain of any symptoms

Patient number	Gender	Age of diagnosis	Length of tosillar descent	Syrinx (yes/no)	Hydrocephalus (yes/no)	Scoliosis (yes/no)	Syndromic/craniosynostosis (yes/no)	Previous CNS infection (yes/no)	GH deficiency (yes/no)	Symptoms	SOS
1n	Female	5	5 mm	Yes	No	No	No	No	Yes	Headache	2.5
2n	Female	13	8 mm	Yes	No	No	No	No	No	Incidental	1
3n	Female	10	5 mm	No	No	No	No	No	No	Incidental	0
4n	Male	7	23 mm	No	No	No	No	No	No	Headache	1
5n	Female	1	11 mm	No	No	No	No	No	No	Incidental	0
6n	Male	13	6 mm	No	No	No	No	No	No	Incidental	0
7n	Male	8	8 mm	No	No	No	No	No	Yes	Incidental	1
8n	Female	12	5 mm	No	No	No	No	No	No	Incidental	0
9n	Male	9	12 mm	No	No	No	No	No	No	Incidental	0
10n	Male	2	8 mm	No	No	No	No	No	No	Incidental	0

brain, spine with, or without CSF flow studies approximately 3 to 6 months after surgery (Fig. 3).

Interestingly, only two patients in the non-operated group and one patient in the operated group presented with headaches as part of their initial diagnosis (Tables 2 and 3). We note that although headache is a common symptom in C1M, however, in children, this has been reported to be less frequent in comparison to the adult population [17, 18]. Next, the use of cine phase-contrast MRI has increasingly been used to evaluate patients with C1M [19]. In order to stay up-to-date with imaging advancements, this additional MRI modality was only recently implemented as part of the radiological assessment. At present, there are four patients in the operated group who underwent this extra scan. Concordantly, we are cognizant that percipient fine-tuning techniques are required to achieve maximal temporal resolution during the course of the MRI scan [19, 20]. As a result, we are working closely with our neuroradiology colleagues for optimal interpretation of our own findings and to correlate them with the patients' treatment outcomes. On a separate note, our curated literature emphasises an association between CNS infection and acquired C1M [15, 21–23]. Theoretically, the formation of post-infective arachnoid adhesions in the CNS is expected to impair CSF circulation, especially so in the presence of foramen magnum obstruction by excess cerebellar tonsillar tissue. This coincides with the extensive inflammatory adhesions/scarring often encountered during surgery in neurosurgical patients with previous CNS infection. An example of such an intra-operative finding was reflected in patient 5o.

Based on these findings, the statistical analyses that generate the current version of our predictive score were inevitably

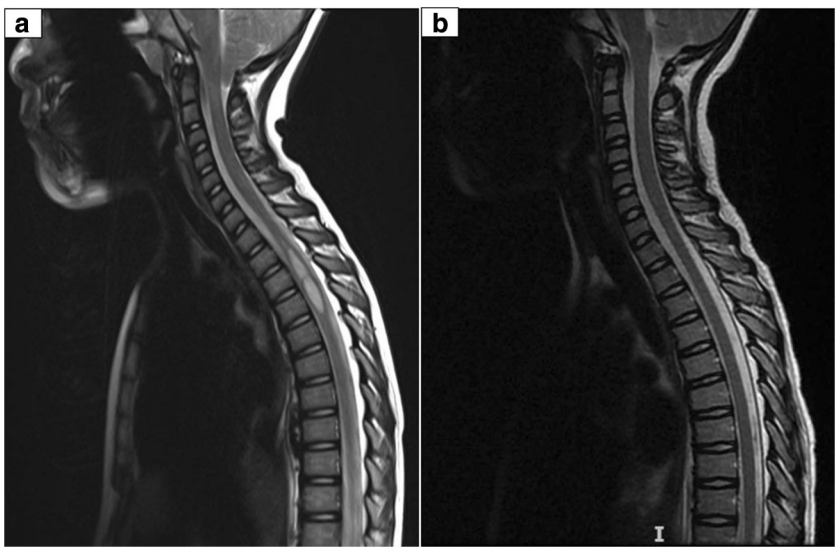

Fig. 2 **a** Representative T2-weighted sagittal MRI image of patient 1n that show protusion of peg-like, cerebellar tonsils below the foramen magnum margins, associated with a lobulated syrinx in the thoracic cord. **b** Corresponding MRI image of the same patient done 36 months later show that the cerebellar tonsils are no longer low-lying and there is interval resolution of the syrinx

Table 3 Table showing list of C1M patients who underwent surgery (hence, labelled as 'o'). Of interest, 2 patients had a SOS of 2 each (patients 10o and 17o). Both cases are discussed in detail in the main manuscript

Patient number	Gender	Age of diagnosis	Length of tonsillar descent	Syrinx (yes/no)	Hydrocephalus (yes/no)	Scoliosis (yes/no)	Syndromis/ craniosynostosis (yes/no)	Previous CNS infection (yes/no)	GH deficiency (yes/no)	Symptoms	SOS	Type of operation	Length of hospital stay
1o	Female	3	6 mm	Yes	Yes	Yes	No	No	No	Progressive scoliosis	3	SDD	7 days
2o	Female	17	10 mm	Yes	No	Yes	Yes(Williams)	No	No	Progressive scoliosis, long tract signs	3	SDD	8 days
3o	Female	12	11 mm	Yes	No	Yes	No	No	No	Progressive scoliosis, long tract signs	3	SDD	9 days
4o	Female	1	9 mm	No	Yes	No	Yes(Beare-Stevenson cutis gyrate)	No	No	Central sleep apnoea	3	SDD	8 days
~	~	5	6 mm	~	~	~	~	~	~	Central sleep apnoea	3	SDD	5 days
5o	Female	13	12 mm	Yes	Yes	Yes	No	Yes	No	Long tract signs	4	SDD	11 days
6o	Male	8	8 mm	Yes	No	No	Yes(Holt-Oram)	No	Yes	Long tract signs	4	SDD	4 days
7o	Male	7	6 mm	Yes	No	Yes	No	No	No	Progressive scoliosis	3	SDD	4 days
8o	Male	14	12 mm	Yes	No	Yes	No	No	No	Long tract signs	3	SDD	5 days
9o	Female	13	10 mm	Yes	No	Yes	No	No	No	Long tract signs	3	SDD	4 days
10o	Male	10	13 mm	Yes	No	No	No (but triplet sister has craniosynostosis)	No	Yes	Incidental	2*	SDD	5 days
11o	Female	5	11 mm	Yes	No	Yes	No	No	No	Cranial nerve palsies	3	SDD	4 days
12o	Female	16	15 mm	Yes	No	Yes	No	No	No	Long tract signs	3	SDD	5 days
13o	Female	8	15 mm	Yes	No	Yes	No	No	No	Progressive scoliosis	3	SDD	3 days
14o	Female	12	10 mm	Yes	No	Yes	Yes(GoldenHar)	No	No	Long tract signs	4	SDD	4 days
15o	Male	4	12 mm	No	Yes	No	Yes(craniosynostosis)	No	No	Central sleep apnoea	3	SDD	10 days
16o	Male	18	11 mm	Yes	No	Yes	No	No	No	Progressive scoliosis	3	SDD	3 days
17o	Female	11	12 mm	No	No	No	No	No	No	Tussive headaches, sleep apnoea	2*	SDD	14 days
18o	Male	5	9 mm	Yes	No	No	No	No	Yes	Long tract signs	3	SDD	4 days
19o	Female	15	7 mm	Yes	No	Yes	No	No	No	Long tract signs	4	SDD	7 days

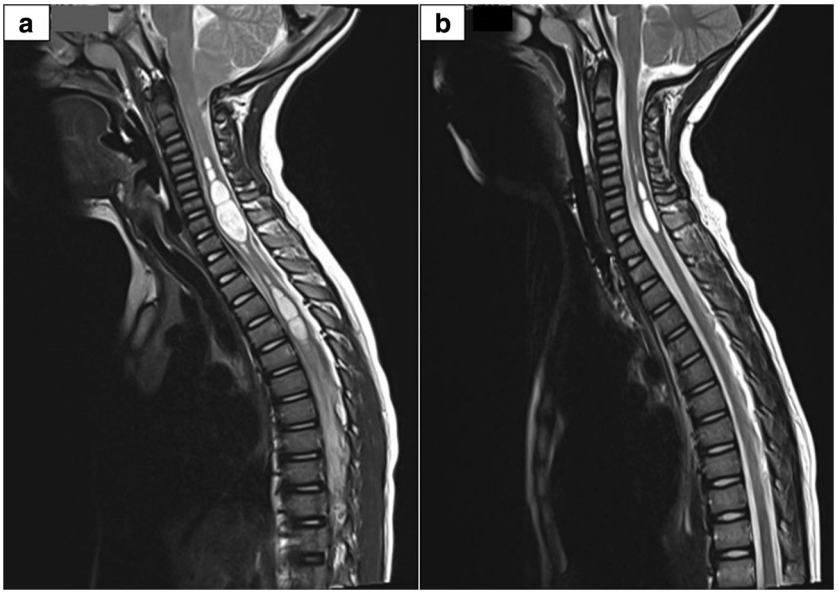

Fig. 3 Representative pre- and post-operative T2-weighted sagittal MRI images of patient 18o depicted in **a**, **b**, respectively. **a** There are low-lying cerebellar tonsils associated with an extensive cervico-thoracic syrinx. **b** Post-surgical decompression, there is unimpeded CSF flow at the level of the foramen magnum with the cerebellar tonsils no longer low-lying and the syrinx significantly smaller

influenced by the data the patients presenting with headache, who underwent cine-phase MRI CSF flow studies and had previous CNS infection, in conjunction with literature correlation. Nevertheless, we reiterate that this is a prospective study in our unit. The SOS is expected to evolve as more patients are recruited; in concordance with progressive, global insights in C1M.

Current reflections and future work

Putting it all together, our study relies on the ongoing inclusion of patient data and conscientious in-house audits to mature our prediction analysis over time. As an institution, there is hope that interval versions of the SOS will pave the way for better management of our paediatric C1M patients, and simultaneously, highlight the differences (if any) between our local cohort of patients and those overseas in the future.

Following this, we are also aware that post-operative outcome assessment for C1M has been reputed to be difficult because of the lack of a reliable scoring system [24]. Recently, the Chicago Chiari Outcome Scale (CCOS) has been validated in the paediatric population [7, 24, 25]. We are reviewing its reliability in our own cohort of our C1M patients. Other aspects of C1M under consideration include posterior fossa volume quantification, evaluation of spinal syrinx characteristics, role of various dura sealants, biological study of tissue adhesions and CSF components encountered during surgery and suitability of other types of imaging, such as a standing MRI. Simultaneously, continuous efforts are made to engage the international medical community to ensure we remain updated of novel advancements in C1M. These interactions are essential for the deliverance of better patient care.

Conclusion

In summary, we report our institutional experience in managing paediatric C1M—a challenging condition denoted by a diversity of opinions. This paper serves to highlight salient points of our practices and is by no means, exhaustive. Meanwhile, we advocate global collaboration, multidisciplinary inputs and ongoing research to reach a practical consensus for affected patients.

Compliance with ethical standards

Conflict of interest We, the authors of this manuscript, report no funding, financial support or industrial affiliations received for the writing of this article. In addition, we report no conflict of interest concerning the material or methods used in this paper. This manuscript has not been published and is not being considered for publication elsewhere.

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