Acta Neurochirurgica (2020) 162:1583–1584
https://doi.org/10.1007/s00701-019-03900-w

EDITORIAL (BY INVITATION) - NEUROSURGERY GENERAL

# Surgery and neuropsychological functions changes in Chiari I malformation: two faces of the same medal?

Massimiliano Visocchi [1]

Received: 12 March 2019 / Accepted: 28 March 2019 / Published online: 21 May 2019


In 1891, Hans Chiari described a group of congenital hindbrain anomalies, which were eventually named after him. He classified these malformations into three types (Chiari malformations I, II and III), and 4 years later added the Chiari IV malformation. Nevertheless, researchers have been encouraged to propose new classifications to encompass some variants not fitting Chiari's original descriptions (e.g. Chiari 0, Chiari 1.5 and Chiari 3.5 malformations). Each has distinct anatomical characteristics and some of these are extremely rare and incompatible with life (e.g. Chiari 3.5). Some physicians might be unfamiliar with the newer classifications of Chiari malformations because these conditions are rare or even unique. In fact, Chiari IV malformation has been further clarified. Furthermore, care is needed in using the term "Chiari IV malformation", which must be consistent with Chiari's original description, i.e. an occipital encephalocele containing supratentorial contents [2].

Arnold Chiari malformation type I (ACM) is a common and often debilitating neurological disease. Efforts to improve treatment of ACM are often impeded by inconsistent and limited methods of evaluating clinical outcomes. To understand current approaches and lay a foundation for future research, some authors have conducted a review of the methods used in original published research articles concluding that, in many clinical studies examined, the outcomes in patients treated for ACM are evaluated according to methods inconsistent and frequently not comparable. From the complications of the efforts to analyse results across studies, the need of validation become proactive to a new deal of more reliable follow-up determination [5].

As a matter of fact, in a neurosurgical population, similarly to hydrocephalus, ACM appears to significantly interfere with behavioural aspects and the Quality-of-life (QoL). Over the last decade, there has been growing evidence that neuropsychological deficits, principally in the executive functions, may be involved in the pathogenesis of ACM. Some studies have been conceived in order to compare changes in cognitive functions in patients with ACM and healthy subjects. From a pure methodological point of view, the neuropsychological profile of these patients has been compared with healthy controls and neuropsychological tests has been administered to both ACM patients and healthy controls in order to assess the frontal executive functions of vigilance or selective attention, mental flexibility, planning, and concept formation. The results obtained has suggested that ACM patients are affected in the processes of inhibition and self-control as well as in attention capacity and maintaining a course of thought and action so providing evidence of possible deficits or anomalies in the cognitive executive functions of these patients [3].

In other publications, patients with ACM have been examined on a large battery of neuropsychological tests, including executive functioning, verbal fluency, spatial cognition, language, verbal memory, processing speed, facial recognition and theory of mind. Results have shown a poorer performance of the clinical group compared with the control group, even after controlling the effect of physical pain and anxious-depressive symptomatology. The findings have suggested the presence of a *generalised cognitive deficit* associated, which makes it necessary to focus attention not only on physical consequences but also on cognitive ones [4].

So QoL changes coexistence, along with the identification of its measurement instruments in ACM management, has become necessary to better understand the severity of the impact on patients' lives. Among them, the Chiari Symptom Profile (CSP) is a valid and reliable instrument designed for this purpose [7].

This article is part of the Topical Collection on *Neurosurgery general*

✉ Massimiliano Visocchi
mvisocchi@hotmail.com

[1] Craniocervical Junction Operative and Research Unit, Institute of Neurosurgery Catholic University of Rome Fondazione Policlinico Universitario A. Gemelli, Rome, Italy

The paper of Almotairi et al. highlights the need to the assess the neuropsychological (NP) functioning, the evaluation of Qu and QoL on adults harbouring ACM since, so far, there is a scarce literature documenting effects of surgery on these aspects [1]. The Hospital Anxiety and Depression scale (HAD), the NP examinations, the self-reported life satisfaction checklist and other health outcome measurements scales (LiSat-11, EuroQol, EQ-5D-5L) have been used [1]. All assessments have been carried out both before and 3 months after surgery. Among the 11 patients who underwent NP assessment, the majority demonstrated cognitive functioning within the normal range. However, postoperatively, their performance in verbal learning, psychomotor speed, colour naming speed, and the ability to manage interference through response selection and inhibition (aspects of executive functioning) was significantly improved. Thirteen patients completed QoL assessments. When LiSat-11 item domains were compared with those of healthy subjects, patients reported a lower level of satisfaction with their life quality both before and after surgery. However, the EQ-5D-5L measurements, i.e. the descriptive system and the visual analogue, indicated that their QoL of life was significantly improved after surgery. This study introduces a new perspective when dealing with ACM by opening new opportunities to widely evaluating pre- and postoperative NP functions and QOL in CMI [1].

The paper opens a wider horizon toward a modern and more effective functional evaluation of postoperative results in such a surgery. Nevertheless, the therapeutic strategy remains a surgery and not a neuropsychological treatment and it needs to be always specified when dealing with a follow-up evaluation.

So far, many surgical strategies on ACM have been conceived so far concerning cranial fossa decompression: anterior (i.e. anterior craniocervical junction compressive pathologies) or a posterior decompression (i.e. crowded posterio cranial fossa). In the case of posterior bone decompression, the dura can be opened totally, partially or simply delaminated. The coagulation of the tonsils can be performed in order to completely remove them or just to produce a shrinkage consistent with a sufficient restoration of CSF cisternal ambiens circulation. The arachnoid membranes lysis can be performed and the IV ventricle opened. A duroplasty can be advisable and many types of dural substitutes include cadaveric, synthetic or autologous [6]. Each procedure interferes with the intracranial system in a different manner and can be associated with different side effects and complications, all driving to the final clinical result, even neuropsychological one.

Each time the authors rule out ACM patients harbouring genetic syndromic conditions associated with behavioural disturbances, they should clarify which one they excluded as well as spend more lines on clinical pictures as Down Syndrome, Mucopolysaccharidosis and other similar conditions. Moreover, they should clarify whether it does exist any relationship between QOL, HAD and hydrocephalus (when associated). According to this issue, also the presence of a cerebrospinal fluid (CSF) fistula (according to the authors, the only complication occurred) should be clearly put in relationship with the presence or not of hydrocephalus.

In other words, ACM is a surgical issue more than neuropsychological and the variable surgical aspects should be more constantly put in relationship with these interesting neuropsychological improvements.

## References


1. Almotairi FS, Hellström P, Skoglund T, Lundgren Nilsson A, Tisell M (2019) Chiari I malformation - neuropsychological functions and quality of life. Acta Neurochir (in press)
2. Azahraa Haddad F, Qaisi I, Joudeh N, Dajani H, Jumah F, Elmashala A, Adeeb N, Chern JJ, Tubbs RS (2018) The newer classifications of the chiari malformations with clarifications: an anatomical review. Clin Anat
3. Besteiro Gonzalez JL, Torres Campa-Santamarina JM (2018) Anomalies in the cognitive-executive functions in patients with Chiari malformation type I. Psicothema 30(3):316–321
4. García M, Lázaro E, López-Paz JF, Martínez O, Pérez M, Berrocoso S, Al-Rashaida M, Amayra I (2018) Cognitive functioning in Chiari malformation type I without posterior fossa surgery. Cerebellum. 17(5):564–574. https://doi.org/10.1007/s12311-018-0940-7
5. Greenberg JK, Milner E, Yarbrough CK, Lipsey K, Piccirillo JF, Smyth MD, Park TS, Limbrick DD (2015) Outcome methods used in clinical studies of Chiari malformation type I: a systematic review. J Neurosurg 122(2):262–272
6. Lavorato L, Spallone A, Visocchi M (2019) Surgical treatment of Chiari malformation in adults: comparison of surgical techniques described in the literature and our experience. Acta Neurochir Suppl 125:139–143. https://doi.org/10.1007/978-3-319-62515-7_20
7. Martinez-Sabater A; Ballestar-Tarin ML; Vazquez-Seoane M; Mari-Avargues L; Saus-Ortega C; Del Carmen Casal-Angulo M (2018) Quality of life in individuals affected by Arnold Chiari malformation: comparison and validation of a measurement instrument. Endocr Metab Immune Disord Drug Targets 18(4):388–396